Calcium pyrophosphate dihydrate

- CPPD - pseudogout
- Crowned dens syndrome: CPPD of atlantooccipital joint causing periodic acute cervicooccipital pain with fever, neck stiffness, and laboratory inflammatory signs
- Etiology
  - unknown etiology
  - In articular, hyaline, and fibrocartilaginous structures
  - Also in joint capsules, tendons, ligaments.
- Age -Usually > 50 years
- Gender – slight female predominance

CPPD

- CPPD is one of many causes of soft tissue calcification (chondrocalcinosis).
- It is not synonymous with chondrocalcinosis and not the only cause of soft tissue calcification.
- Where crystal deposition causes acute clinical manifestation, the term pseudogout should be used.
- Pyrophosphate arthropathy is a term that describes arthropathy secondary to CPPD deposition.
- However, it is often used indiscriminately to refer to chondrocalcinosis too.

Etiology

- Idiopathic
- hereditary
  - AD pattern; mutation in gene ANKH encoding transmembrane inorganic pyrophosphate transporter
- secondary
  - haemochromatosis
  - hyperparathyroidism
  - hypothyroidism
  - hypomagnesaemia [2]
  - previous joint injury
  - ochronosis

Epidimiolgy

- At least 5% of elderly have CPPD at some location in autopsy series
- Knee chondrocalcinosis due to CPPD deposition is found in 9.6% of individuals > 50 years old
- Prevalence of CPPD unknown but increases with age
  - Rate of 45% among people ≥ 85 years
  - Overall population frequency of 1 case per 1,000 persons
- CPPD previously thought to be rare in spine
  - Based on CT, not uncommon

Chondrocalcinosis

- Means visible calcification of both Hyaline cartilage and fibrocartilage.
- Etiology
  - hypercalcaemia, especially hyperparathyroidism
  - arthritides
  - gout
  - Wilson disease
  - haemochromatosis
  - ochronosis
  - trauma: focal chondrocalcinosis in a traumatised joint
  - hypomagnesaemia
  - hypothyroidism
  - oxalosis
  - acromegaly
  - HADD
  - osteoarthritis

Check List

- **Consider**
  - CPPD may be present in axial skeleton without peripheral involvement
  - Diagnosis may be missed at surgery unless surgeon is alerted to possibility of CPPD from imaging
    - Surgical specimens usually sent to pathology in formalin
    - Precaution with surgical specimen as formalin dissolves crystals
- **Image Interpretation Pearls**
  - CT is useful in distinguishing from rheumatoid arthritis, which does not calcify

Imaging

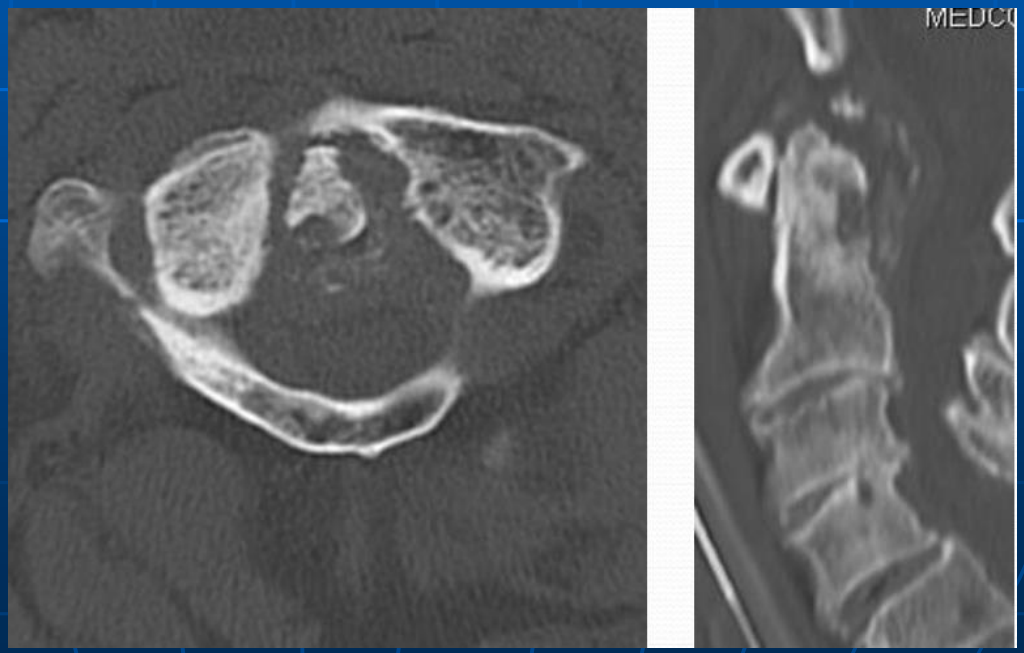
- Spine involvement often seen in absence of peripheral skeleton involvement
- Soft tissue calcifications
  - Usually linear, occasionally globular
  - Seen in ligaments, discs, facet joint capsules, hyaline cartilage
  - Horseshoe-shaped calcification around dens
- Erosions of odontoid process, vertebral endplates
  - Usually sharply demarcated, often corticated
- MR findings are nonspecific
  - Calcium is usually not visible; low signal intensity on all sequences
  - Soft tissue mass surrounding dens
  - Erosions of dens and vertebral endplates

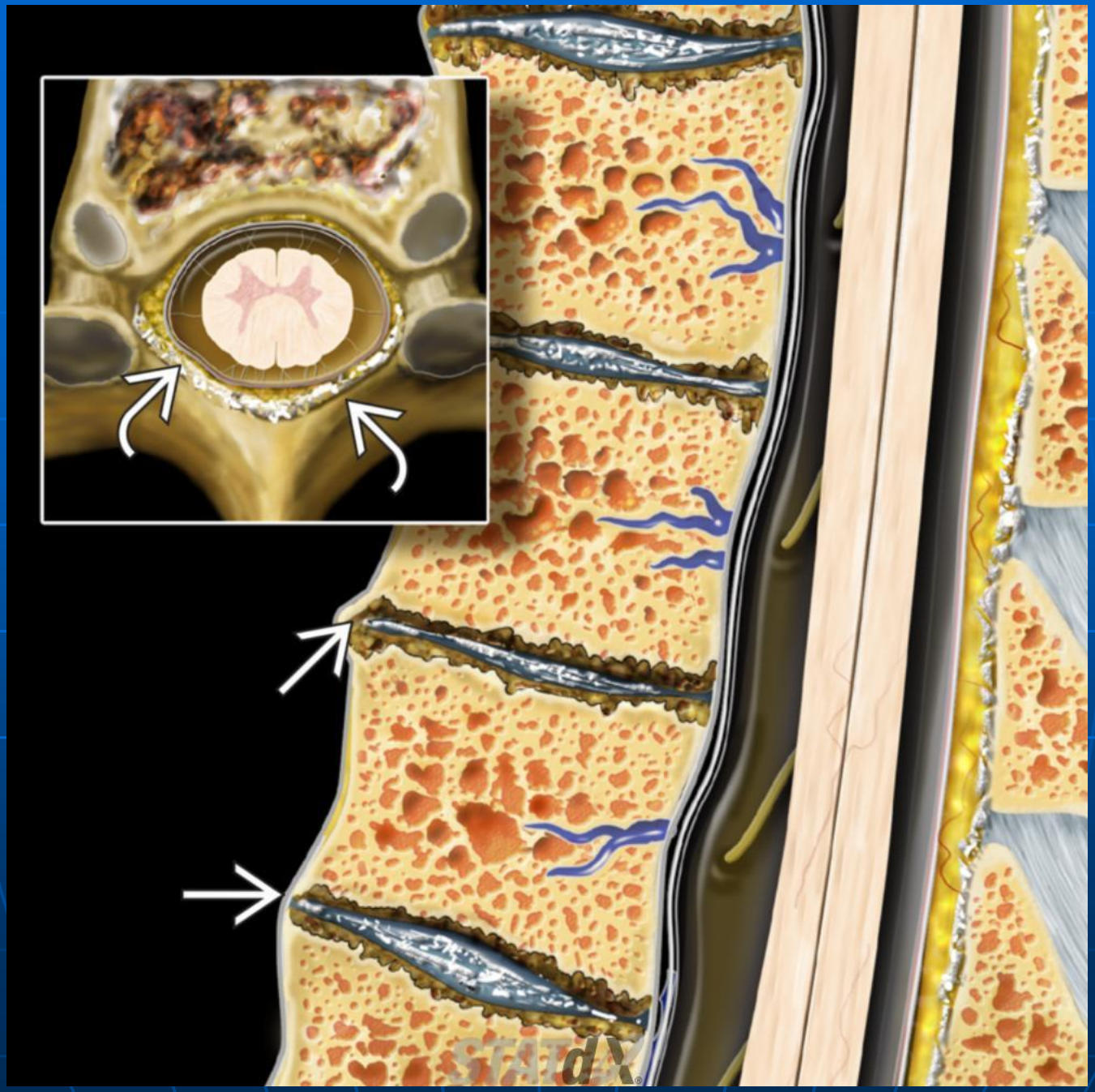
Sagittal CT depicts a retroodontoid soft tissue mass with calcification (white curved arrow) and erosion along the posterior margin of the odontoid process (white solid arrow). Corticated erosions along the vertebral endplate (white open arrow) are also observed. In the setting of acute neck pain and limited range of motion, this represents crowned dens syndrome.

Sagittal T1WI MR (left) demonstrates a lobulated T1-isointense mass posterior to the dens (white solid arrow). The lesion is heterogeneous on the sagittal T2WI MR (right) with areas of hypointensity (white curved arrow). CPPD crystals are often deposited in a fibrocartilaginous matrix and may be located around and within the transverse ligament. The cervical cord is severely compressed (white open arrow).

CPPD Arthropathy

- C1-C2 calcs, (“crowned dens”), soft tissue mass
  - Transverse ligament, sometimes ligament flava
  - Can erode dens similar to RA





Sagittal graphic shows crystal deposition and degeneration of the intervertebral disc (white solid arrow). Calcification of the ligamentum flavum (white curved arrow) is seen on the axial inset graphic.

DDx:

- Degenerative Disc Disease
  - Disc bulges and herniations may calcify when chronic
  - Mild irregularity of vertebral endplate often occurs
    - Generally less severe than erosions of CPPD
- Pseudopannus
  - Thickening of tectorial membrane due to degenerative changes
  - Causes soft tissue fullness around dens
  - May be seen with abnormal motion secondary to more inferior cervical fusion.
- Hydroxyapatite Deposition Disease
  - Rounded calcific deposits in soft tissues
    - Homogeneous, amorphous densities
    - Tendon insertions
    - Periarticular/Intraarticular deposition of hydroxyapatite crystals
      - May involve matrix vesicles and local dysregulation of extracellular PPi homeostasis (PPi: Inhibitor of apatite crystal nucleation)
  - Uncommon in spine
    - Common in shoulder regions

DDx:

- Rheumatoid Arthritis, Adult
  - Odontoid erosions; soft tissue mass may appear identical to CPPD on MR
    - Absence of calcifications distinguishes from CPPD on CT and radiographs
  - Spares intervertebral discs
  - Look for subaxial erosive changes
- Seronegative Spondyloarthropathy
  - Ligament ossifications
  - May appear identical to CPPD on MR
- Pyogenic Osteomyelitis
  - Usually affects only 1 level
  - High signal intensity in disc on T2WI and STIR
  - Phlegmon or abscess often present in spinal canal or prevertebral soft tissues
  - Infected areas show enhancement with gadolinium

DDx:

- Hemodialysis Arthropathy
  - Endplate erosions
  - Calcifications due to hydroxyapatite deposition
- Hyperparathyroidism
  - Osteopenia, endplate erosions, soft tissue calcifications
  - Laboratory values most useful to distinguish from CPPD
- Ochronosis
  - Diffuse disc calcifications and degeneration
- Gout
  - Much less common in spine than CPPD
  - Endplate erosions, soft tissue masses
  - Calcifications are usually minimal
  - Focal "punched out" lesions are typical