Primary Synovial Chondromatosis of the Wrist

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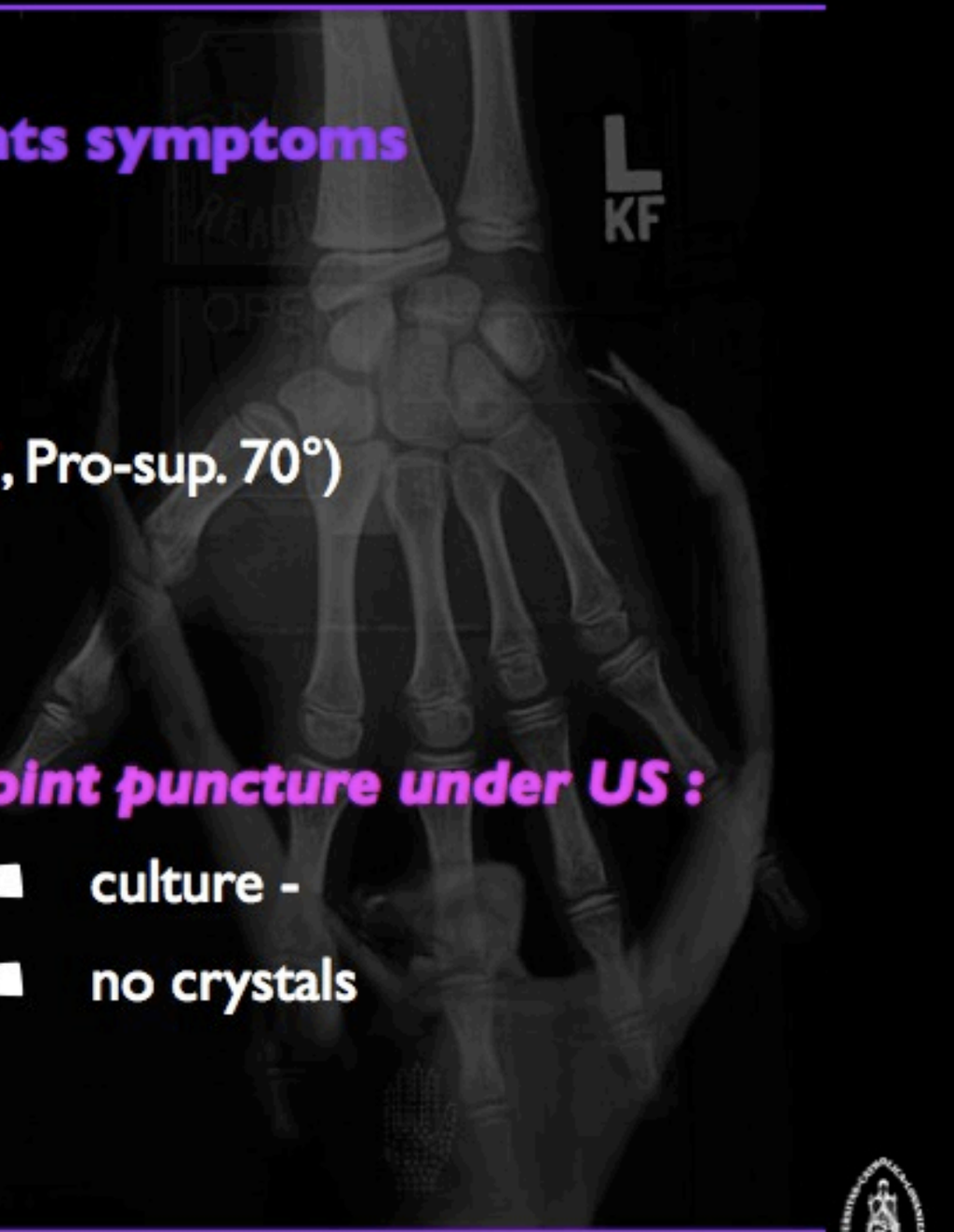
INTRODUCTION

- Primary synovial chondromatosis
  = an unfrequent pathology
- The wrist (intra-carpal site)
  = an extremely uncommon affected site
- Only case seen in our Department
  in Department of Radiology

CASE REPORT

- **CLINICAL CHARACTERISTIC : joints symptoms**
  - 44 y-old, Taxi-man driver
  - > 1y swelling and left wristpain
  - Progressive ↘ ROM (Flex. 75°, Ext. 35°, Pro-sup. 70°)
  - Locking sensation
  - Palpable mass (ulnar side first)

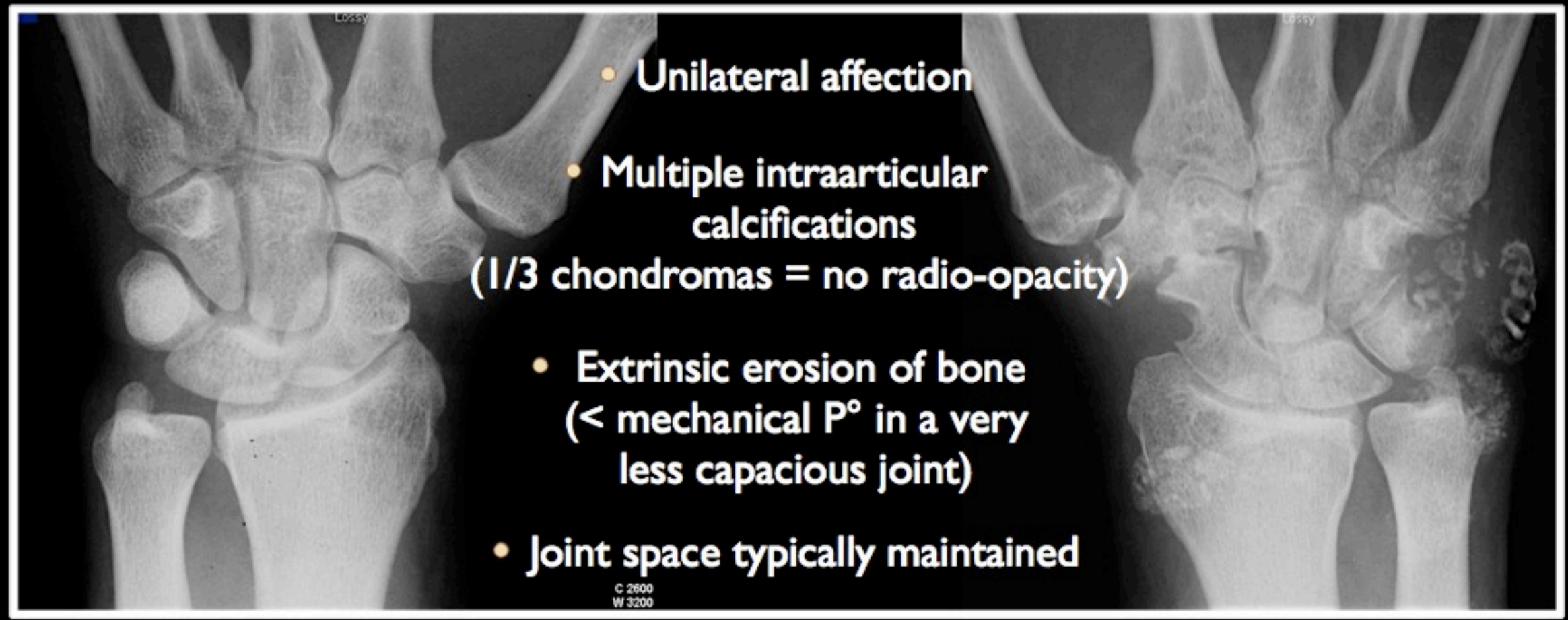


- ***Laboratory tests :***
  - no inflammatory syndrome
  - rheumatoid test -
  - no hyperuricemia
- ***Joint puncture under US :***
  - culture -
  - no crystals

RADIOLOGIC FINDINGS : pathognomonic

Standart X-Ray

AP view

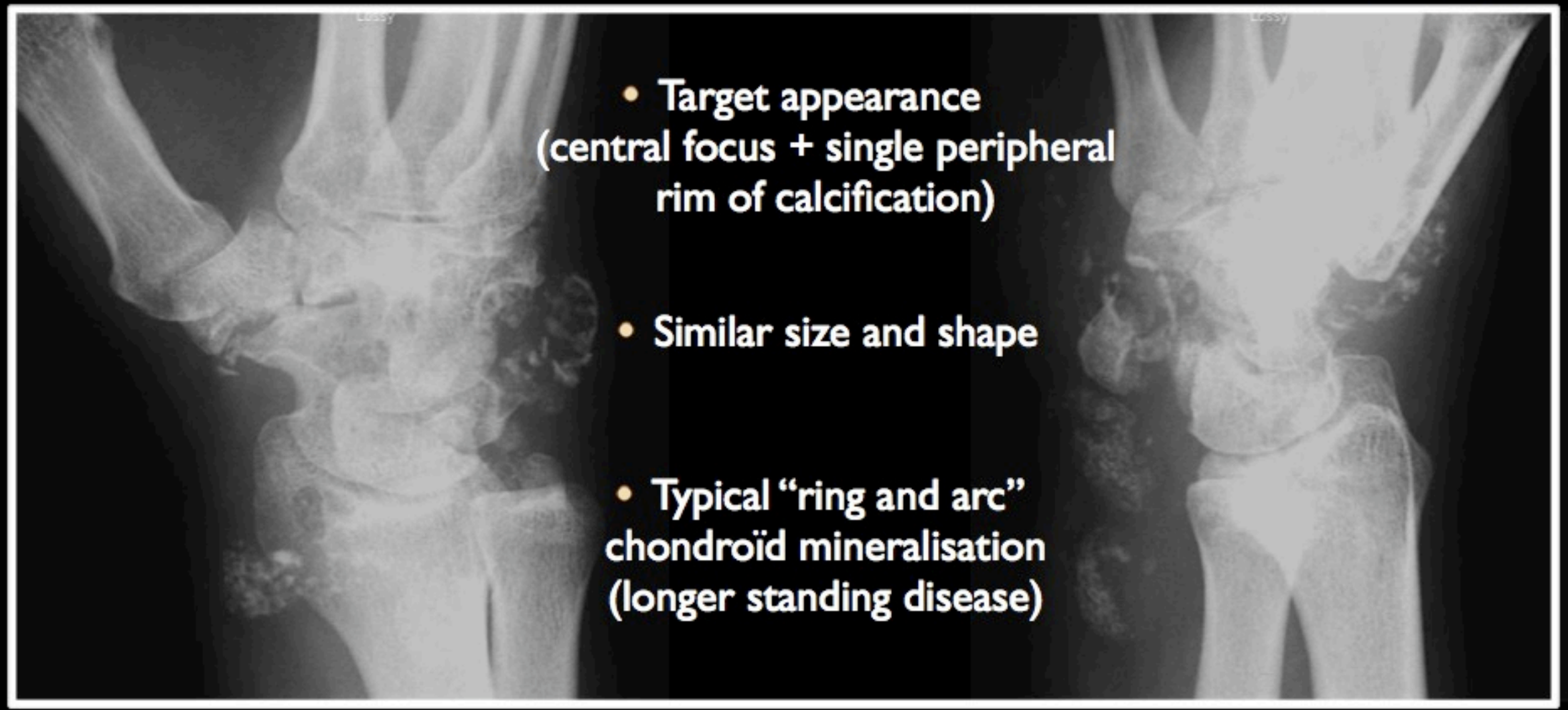


Murphey MD, Vidal JA, Fanburg-Smith JC, Gajewski DA. Imaging of synovial chondromatosis with radiologic-pathologic correlation. Radiographics. 2007; 27(5) : 1465-88

RADIOLOGIC FINDINGS : pathognomonic

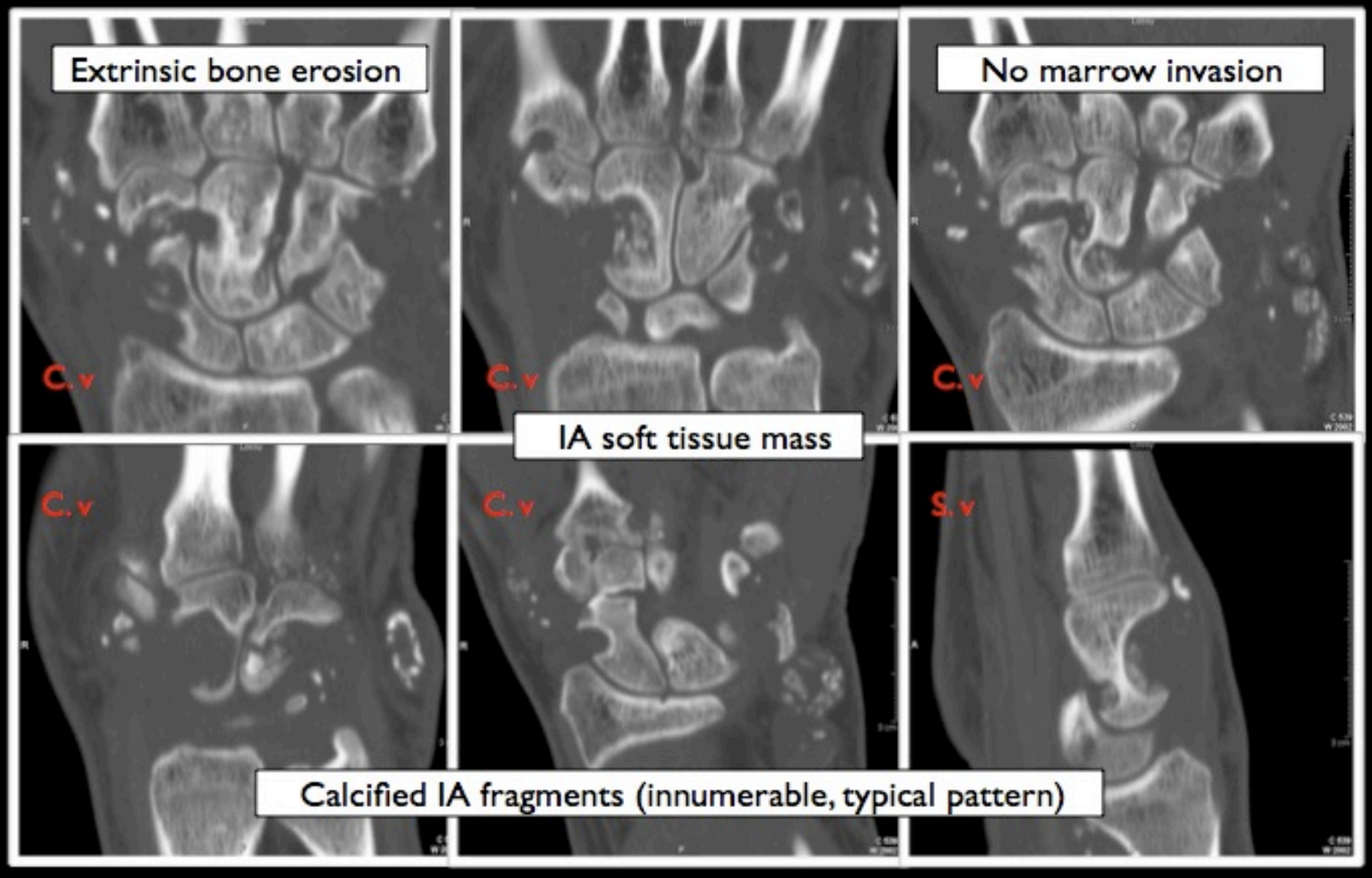
- Standart X-Ray

Oblique incidence



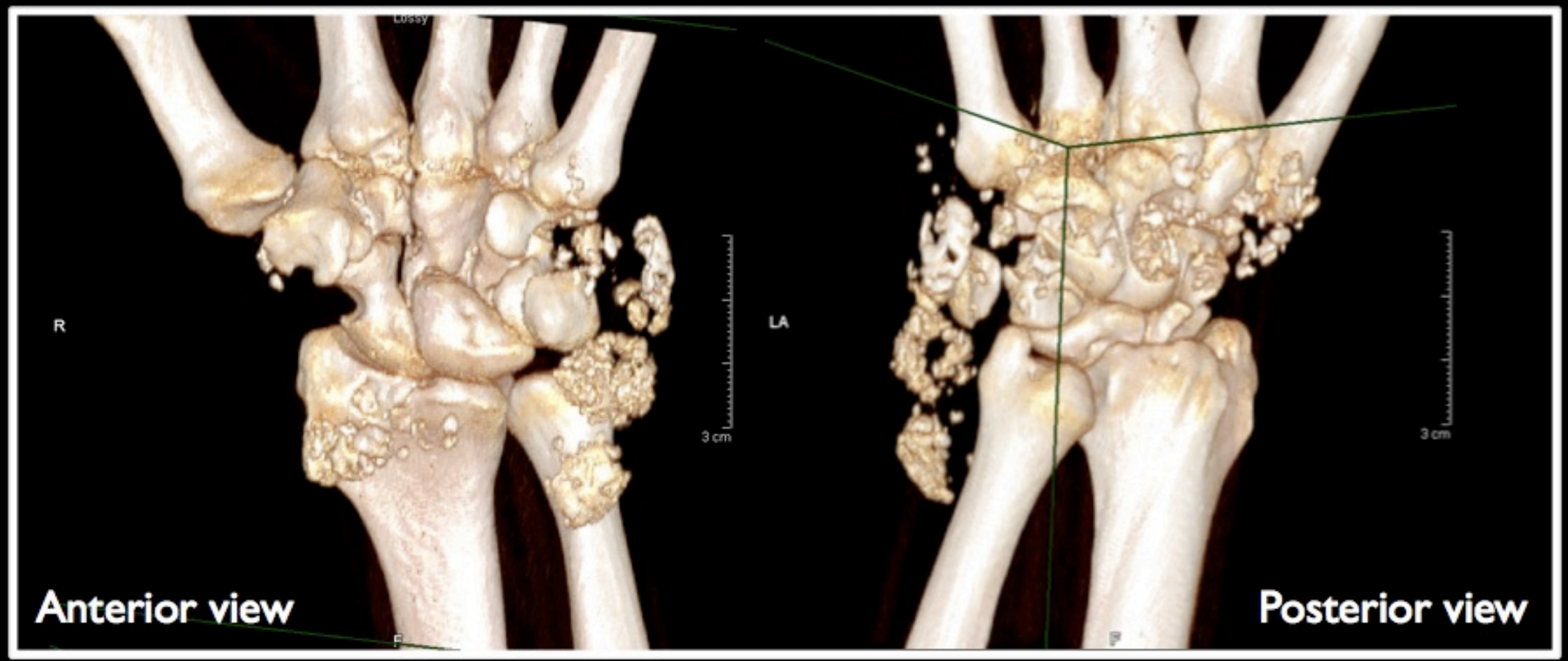
RADIOLOGIC FINDINGS : pathognomonic

- CT : detect and characterize calcification



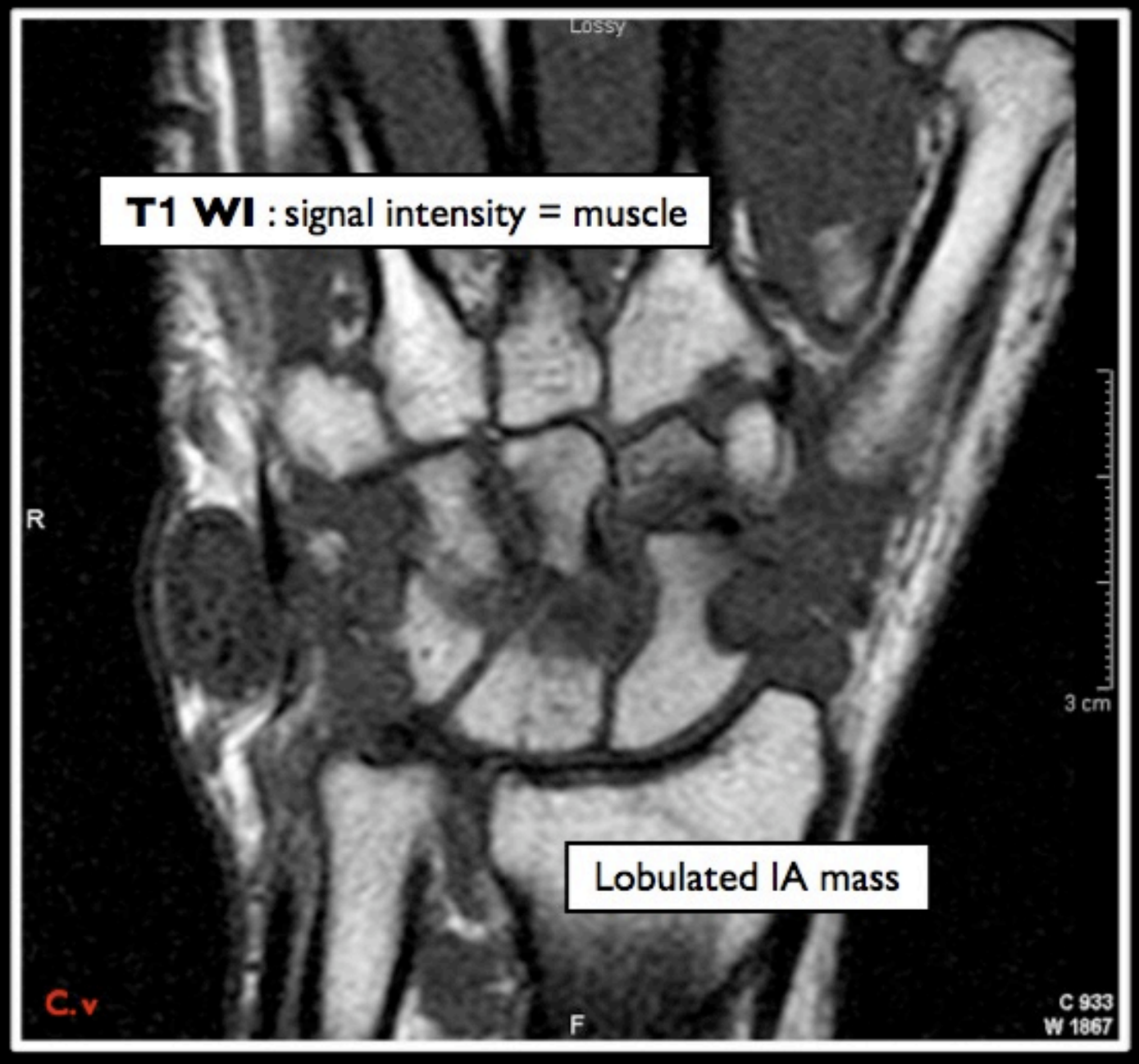
- **RADIOLOGIC FINDINGS : pathognomonic**
  - CT : detect and characterize calcification

3D reconstruction



RADIOLOGIC FINDINGS : pathognomonic

- MRI : marrow invasion ?



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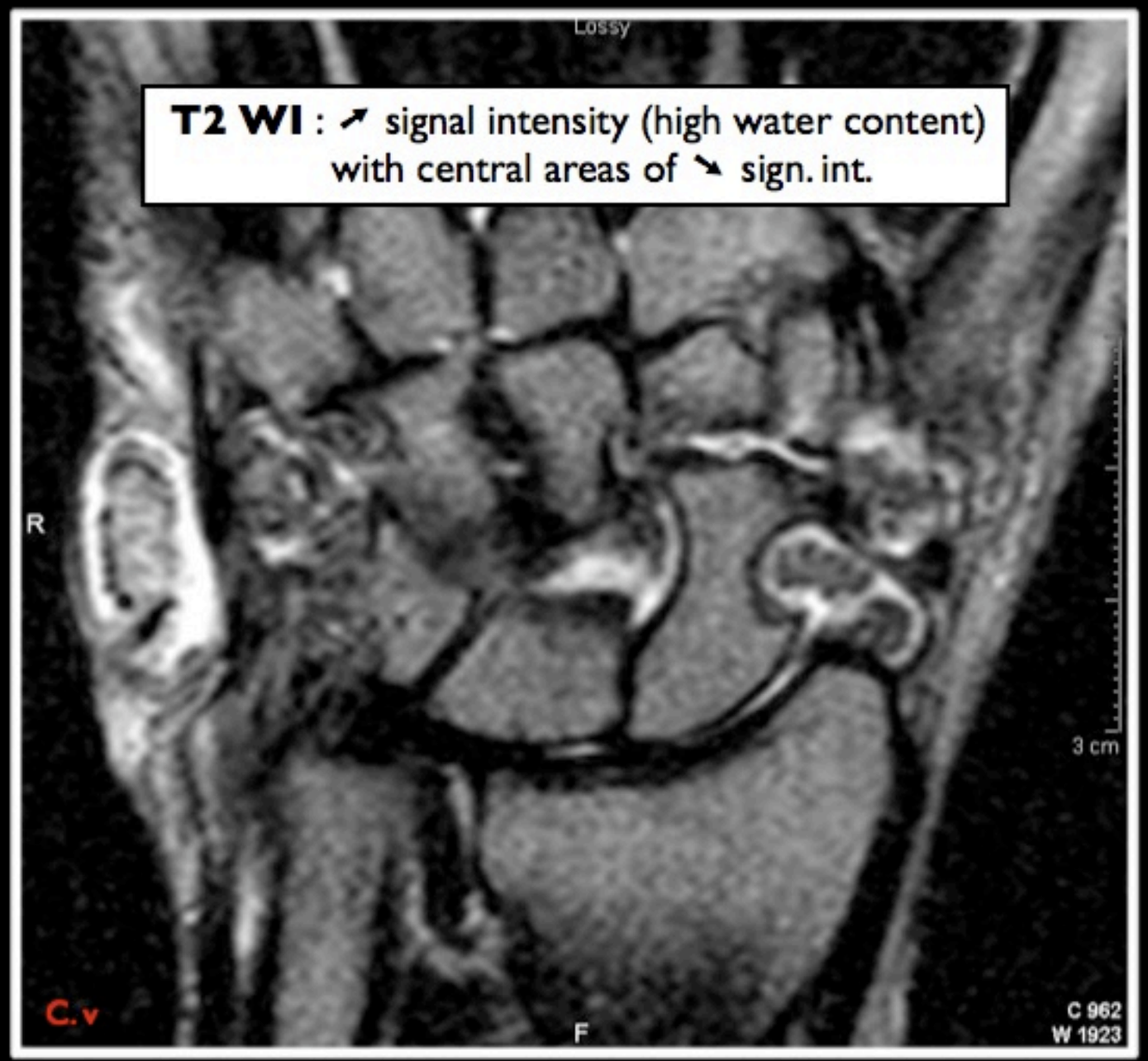

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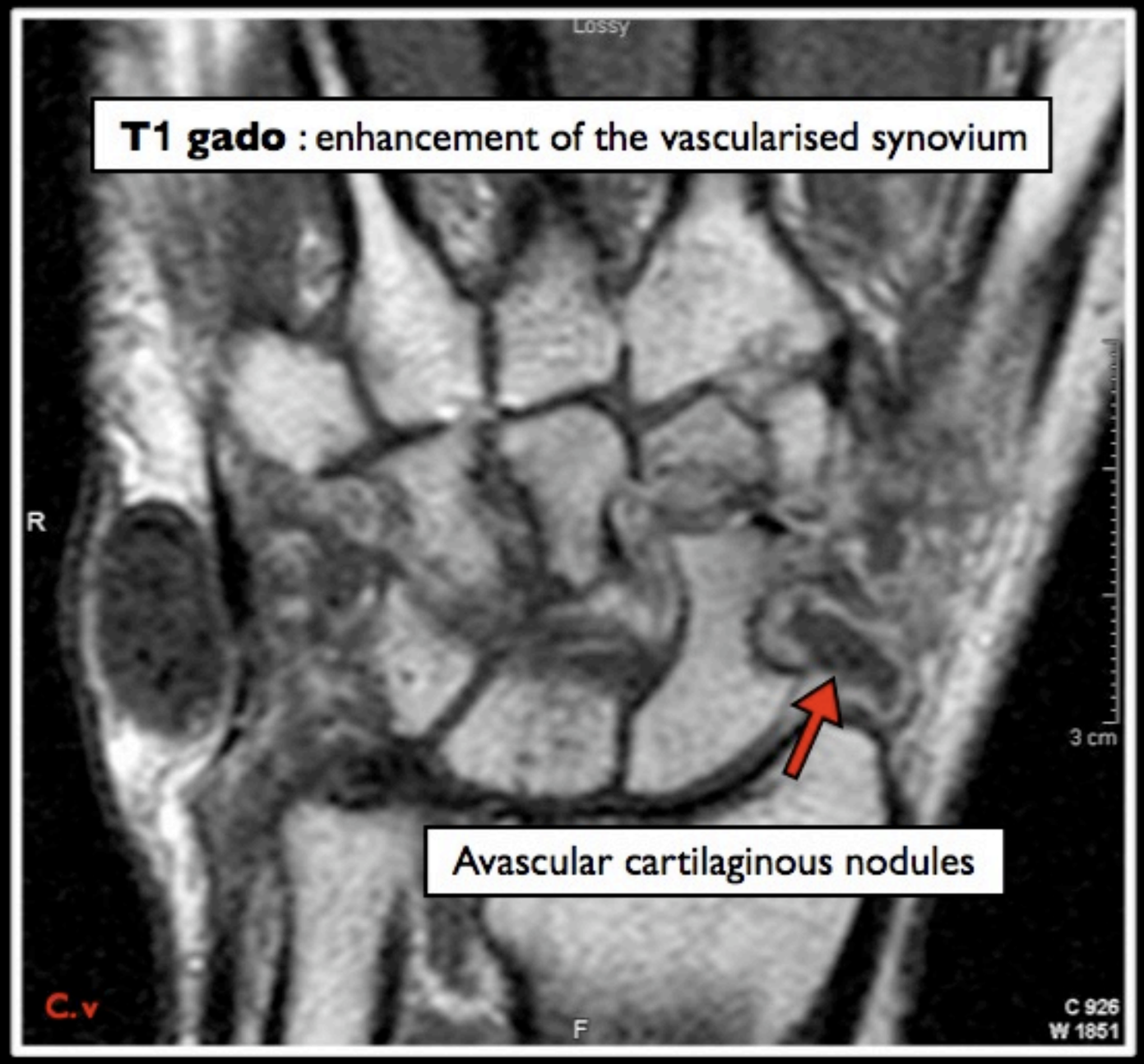
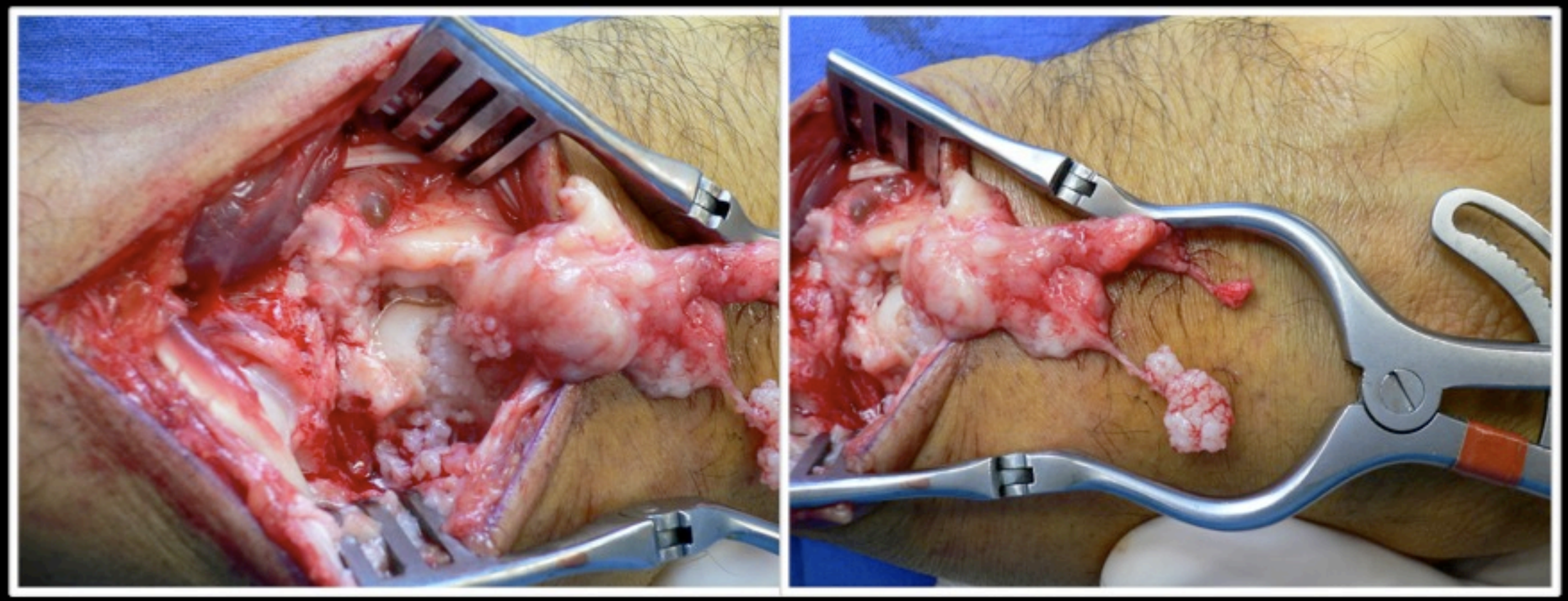


SURGICAL TREATMENT

- Intra-operative views : Synovectomy with Removal of chondral bodies



Posterior approach

SURGICAL TREATMENT

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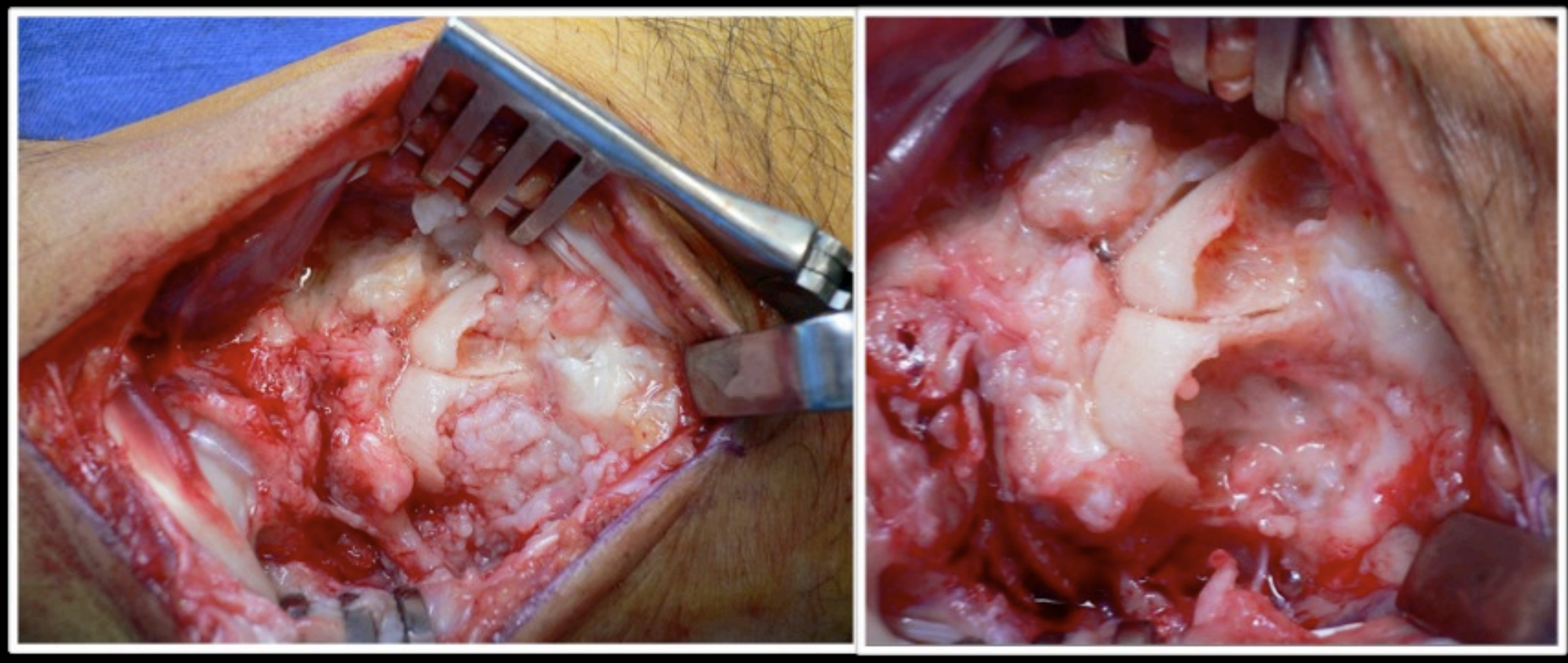

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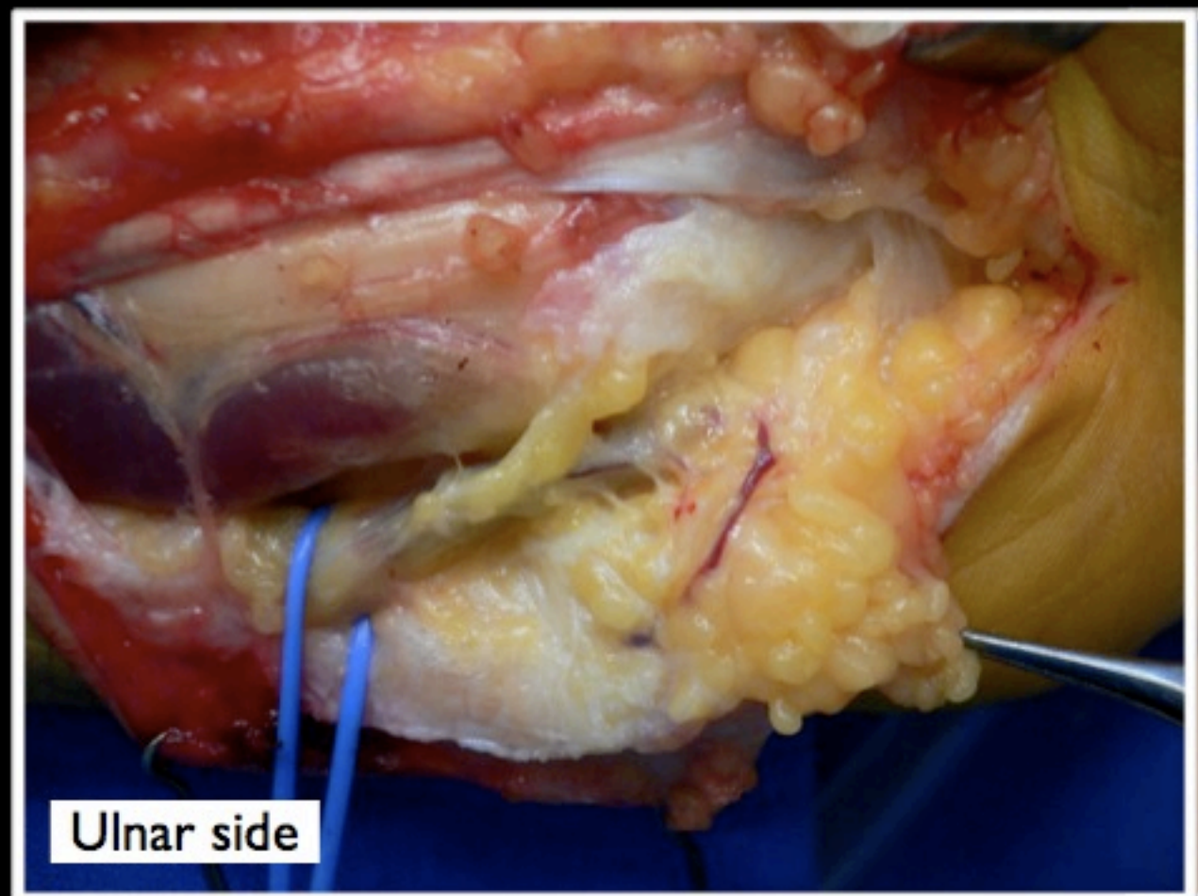
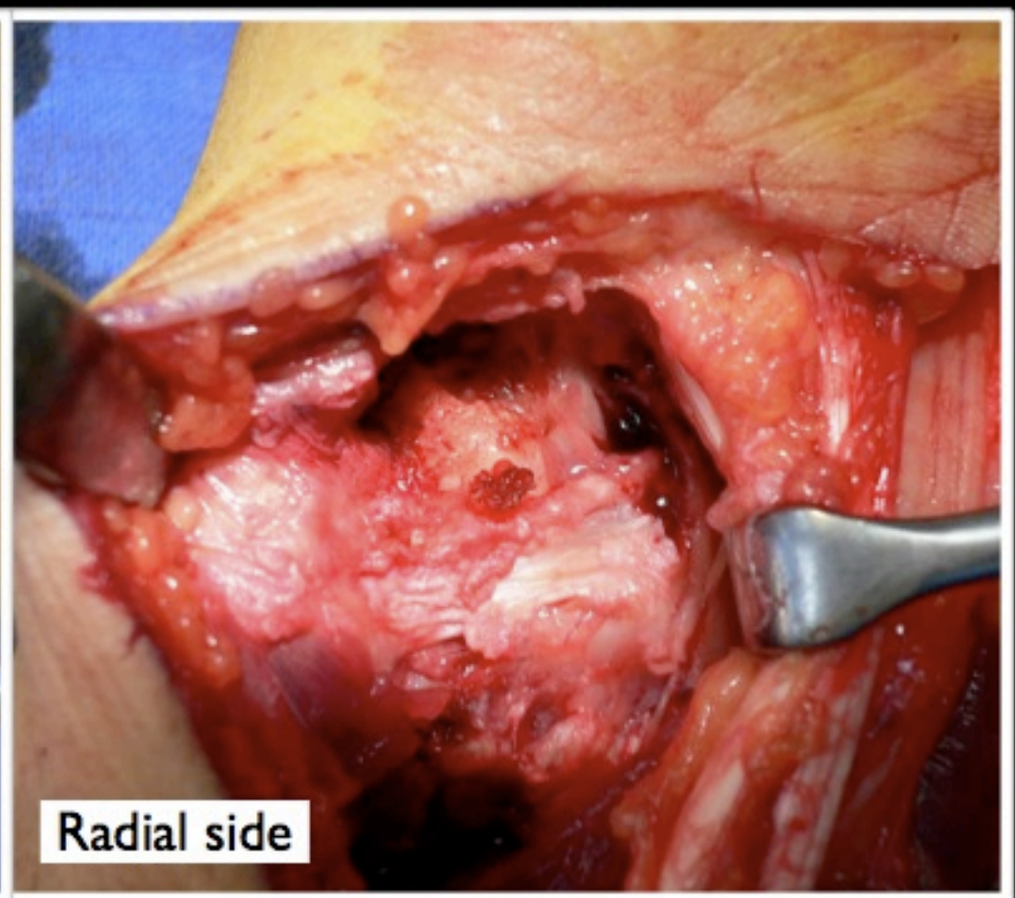
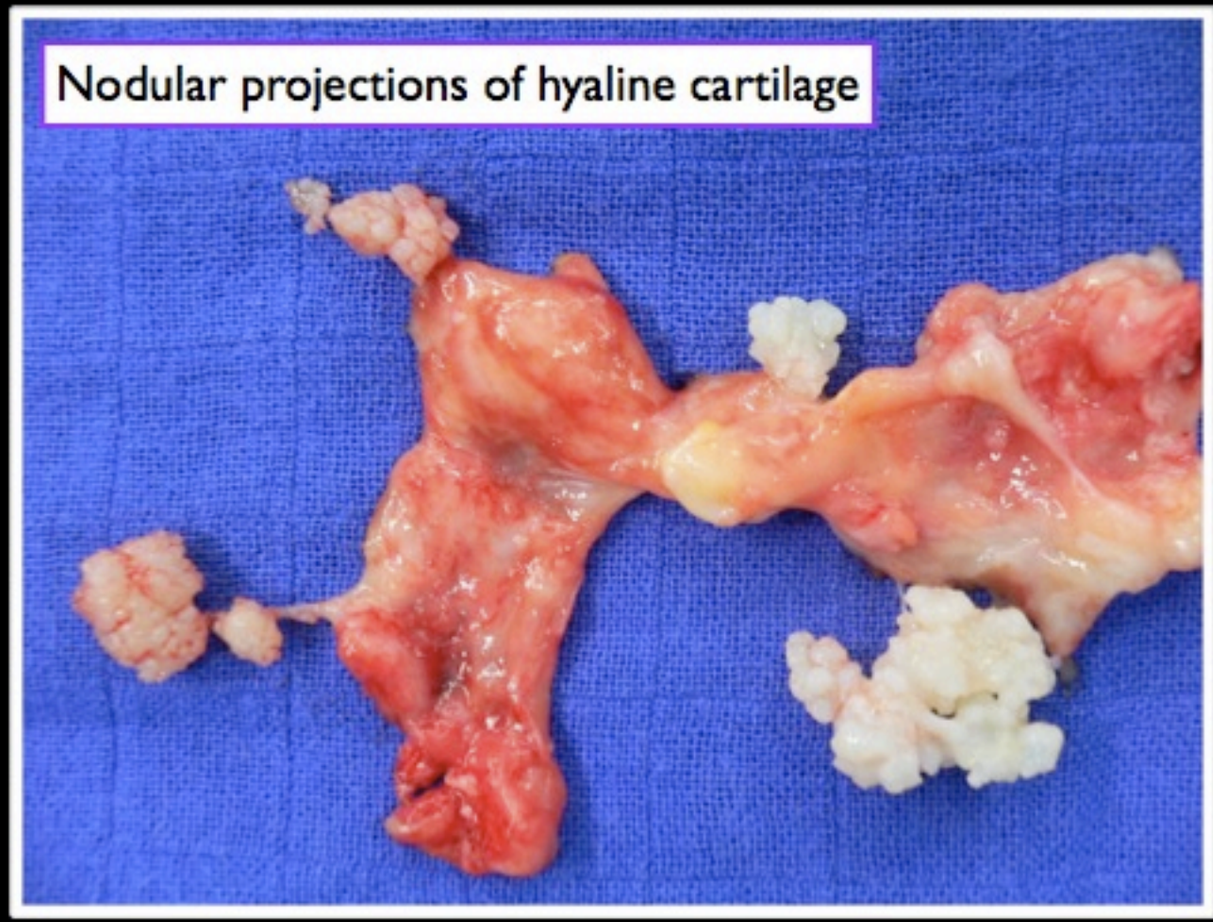




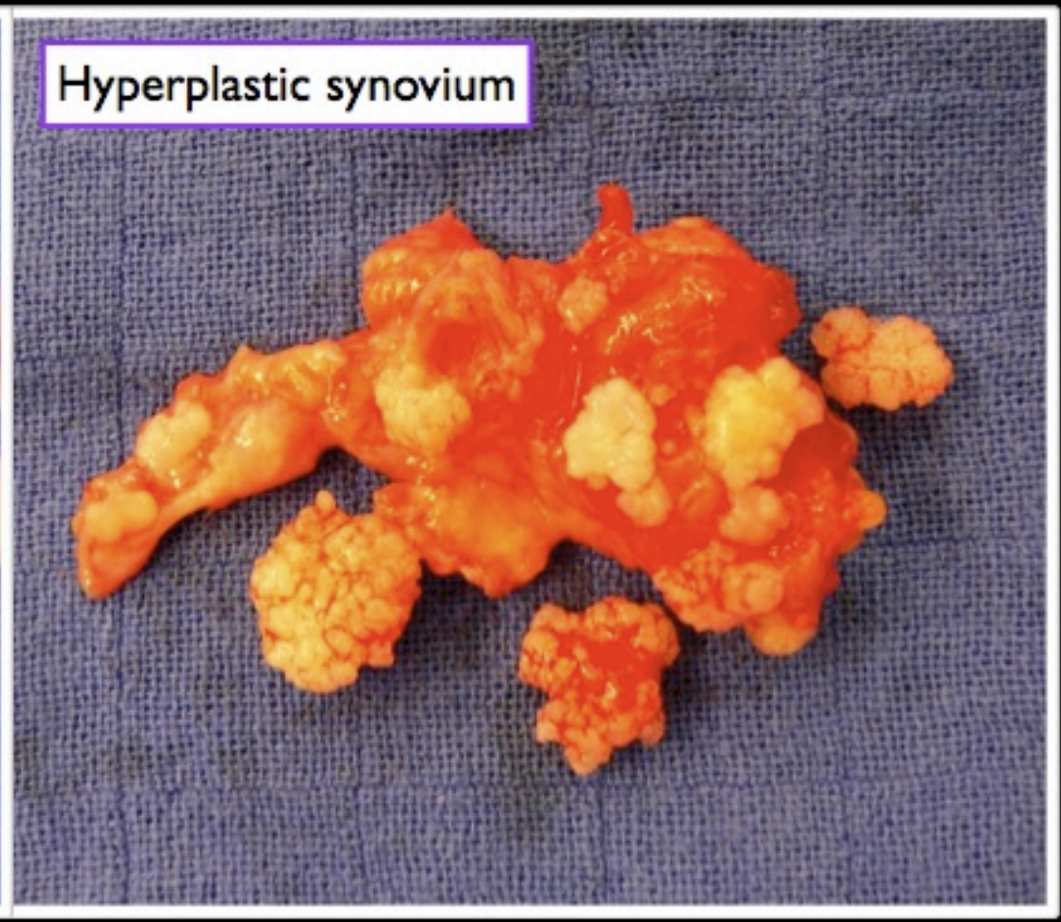
Anterior approach

PATHOLOGIC FEATURES

- Gross pathologic appearance / macroscopic aspect



Posterior synovectomy



Anterior synovectomy

L. Sviland and A.J. Malcom. Synovial chondromatosis presenting as painless soft tissue mass-a report of 19 cases. Histopathology. 1995; 27(3) : 275-279

Murphey MD, Vidal JA, Fanburg-Smith JC, Gajewski DA. Imaging of synovial chondromatosis with radiologic-pathologic correlation. Radiographics. 2007; 27(5) : 1465-88

PATHOLOGIC FEATURES

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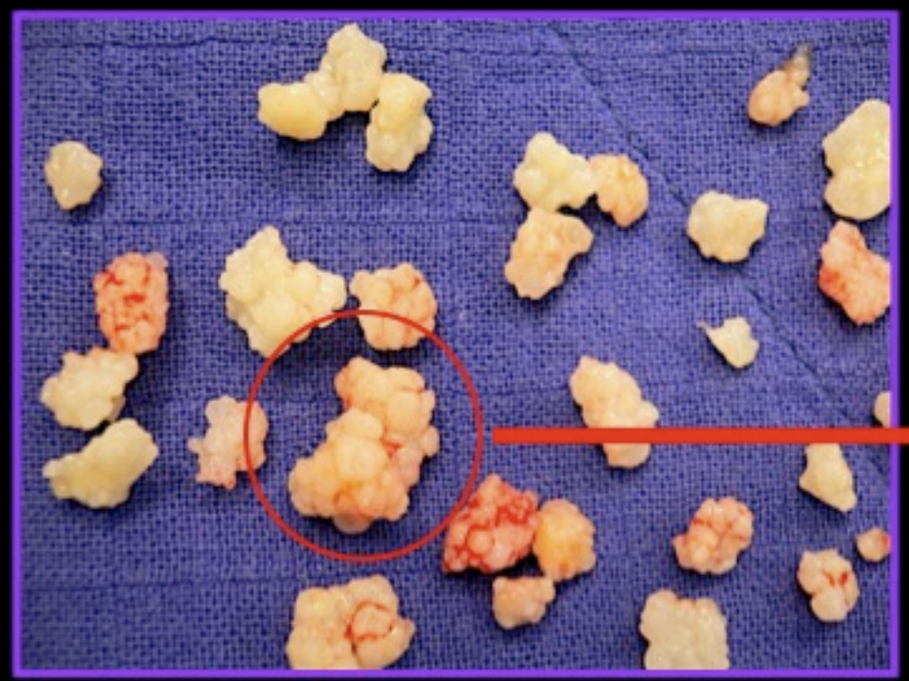


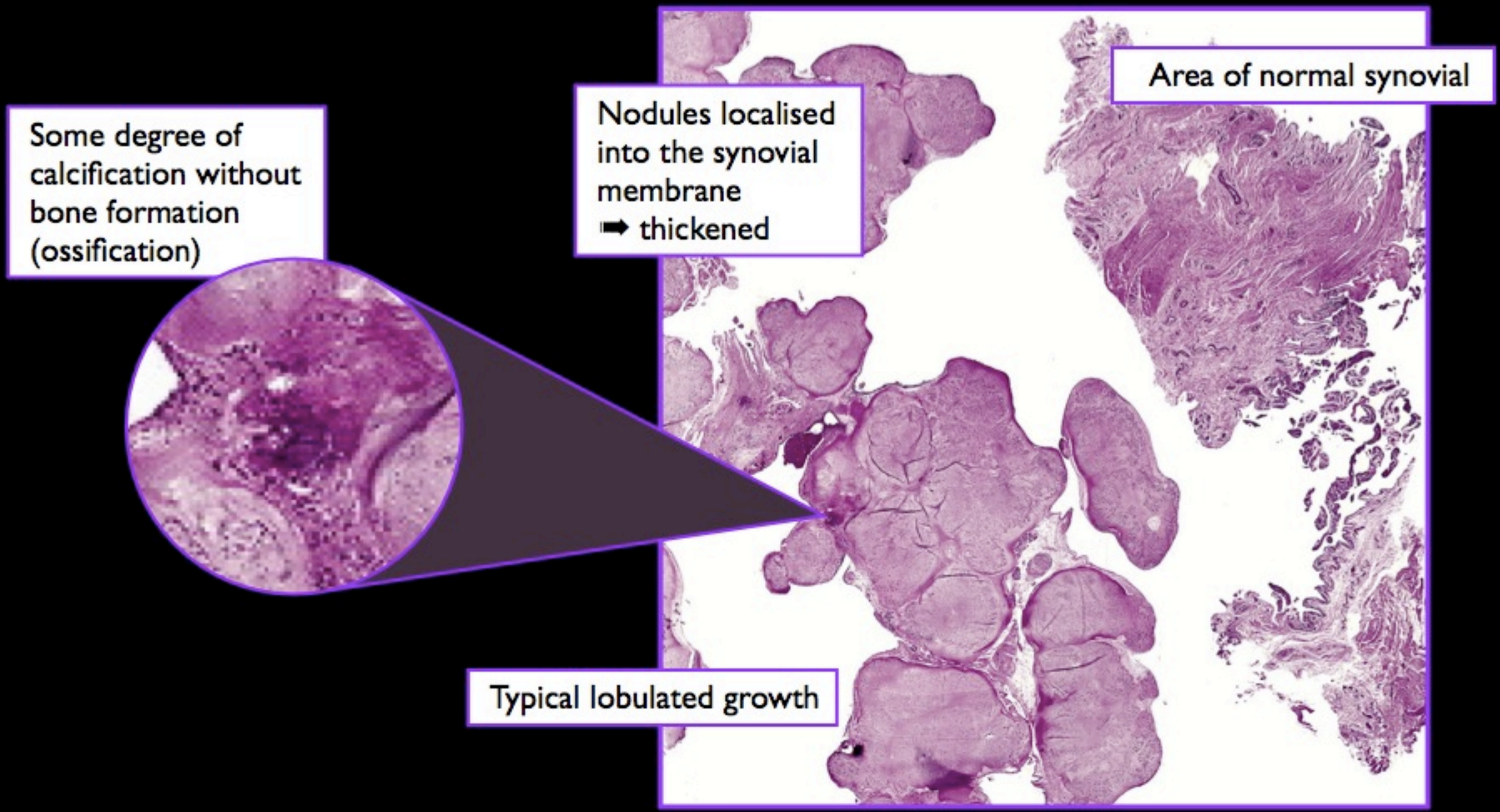
Loose IA chondral bodies

Usually similar in size and shape

Multilobulated typical lesion

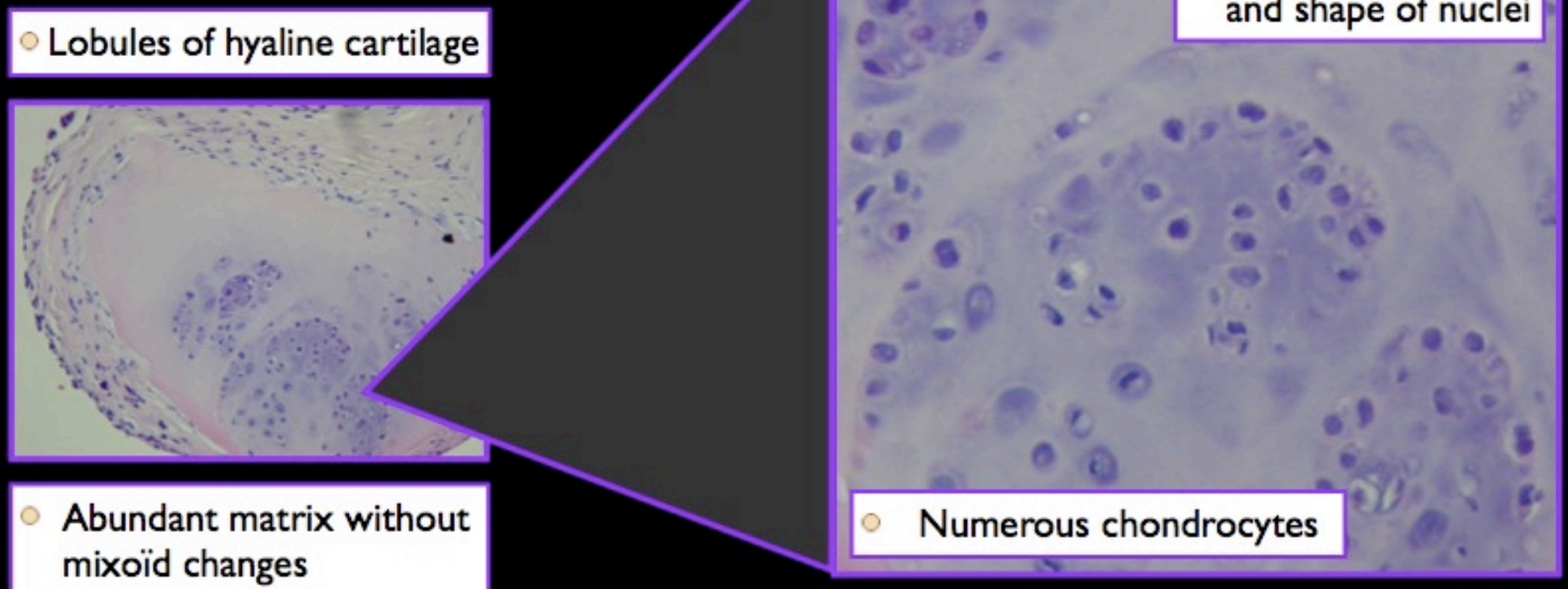
PATHOLOGIC FEATURES

- Microscopic analysis : photograph of the whole-mounted specimen



PATHOLOGIC FEATURES

- Microscopic analysis : high-power photomicrograph



➡ suggest a grade 1 or 2 Chondrosarcoma
Hypercellularity and Nuclear Atypia are TYPICAL of this benign disease

Histological analysis CONFIRMED DIAGNOSIS of PSC

DISCUSSION

- **DEFINITION**
  - uncommon benign process
  - etiology : - in the past = originally considered a chondroïd metaplasia in the synovium ➡ IA chondral bodies

    - actually = cytogenetic evaluation ➡ molecular abnormalities (expression of FGF 2-3, proto-oncogene C-ERBB2, chrom 6 abnormalities, ...)

Hopyans S,, NAdesan P,Yu C,Wunder J,Alman BA. Dysregulation of hedgehog signalling predisposes to synovial chondromatosis. J Pathol 2005; 206 : 143-150.

Robinson D, Hasharoni A, Evron Z, Segal M, Nevo Z. Synovial chondromatosis : the possible role of FGF receptor 3 in its pathology. Int J Exp Pathol 2000; 81: 183-189.

Hocking R, Negrine J. Primary synovial chondromatosis of the subtalar joint affecting two brothers. Foot Ankle Int 2003; 24 : 865-867.

DISCUSSION

DEFINITION

Sato J, Segami N, Suzuki T, Yoshitake Y, Nishikawa K. The expression of fibroblasr growth factor - 2 and fibroblast growth factor receptor - 1 in chondrocytes in synovial chondromatosis of the temporomandibular joint : report of 2 cases. Int J Oral Maxillofac Surg 2002; 31 : 532-536.

Buddingh EP, Krallman P, Neff JR, Nelson M, Liu J, Bridge JA. Chromosome 6 abnormalities are recurrent in synovial chondromatosis. Cancer Genet Cytogenet 2003; 140 : 18-22.

➡ BENIGN NEOPLASTIC process more than METAPLASTIC process

DISCUSSION

- **DEFINITION**
  - affects predominantly ♂ (2-4x more than ♀)
  - 3rd to 5th decades of life
  - commonly occurs within joints, rare cases in EA sites (tendon sheath and bursa)
  - invariably a mono-articular disease (in large series)
  - knee = the most commonly affected site

    Follow by : hip, elbow, shoulder, ankle

    Less commonly : MP, IP, distal RU, AC, TM, proximal T-F joints

Intra-carpal site NOT DESCRIBED in the literature.

DISCUSSION

- **DEFINITION**
  - symptoms : - joint symptoms usually (pain, swelling, locking, instability, ...)
    - insidious and gradually progressive
    - duration before diagnosis +/- 5y
  - peripheral enchondral ossification in long-standing disease
    BUT histologically absent in 45% of cases (Davis' study 1998)

➡ Synovial Osteochondromatosis is NOT A GOOD TERM

DISCUSSION

- **DIFFERENTIAL DIAGNOSIS : similar complaints**
  - Secondary synovial chondromatosis :

    - very common abnormality

    - older patients

    - joint abnormalities (mechanical or arthritic conditions)

    - most commonly affect : knee, hip, shoulder

    ➡ Radiologically : - underlying articular disease

    - smaller number and variable size of chondral fragments

    ➡ Pathologically : concentric ring of growth (several ring of calcification)

DISCUSSION

- **DIFFERENTIAL DIAGNOSIS : similar complaints**
  - Chondrosarcoma :
    - arise in bone and extend into the joint
    - ➡ Radiologically : can be distinguished
    - ➡ Pathologically : similar histologic atypia
  - Soft-tissue chondroma :
    - ➡ location : - hand and feet
      - no IA

DISCUSSION

- **TREATMENT**

Optimal R/ = Synovectomy and removal of the chondral bodies

... BUT CONTROVERSY EXISTS....

SHPITZER 1990 and MAURICE 1988 studies : no difference in prognosis

OGILVIE - HARRIS study 1994 : 60% recurrence rate removal chondral bodies alone

DISCUSSION

- **EVOLUTION-PROGNOSIS**
  - recurrence rate 3-23% (related to incomplete resection in many cases)
  - chronic disease ➡ secondary OA
  - malignant transformation to chondrosarcoma :
    - extremely rare event (5% of cases)
    - signs of malignancy : - multiple local recurrences
      - rapid ↗ of the lesion
      - rapid clinical deterioration
      - marrow invasion

➡ R/ Amputation

CONCLUSION

- Benign self-limited process, may recur locally
- Histologic appearance suggest a chondrosarcoma

➡ Histologic CORRELATION with the Radiologic appearance

= ESSENTIAL for a correct diagnosis and treatment.

Thank you