Sarcomes des tissus mous

Dr B. Morattel

27.04.2021

Sarcomes: généralités

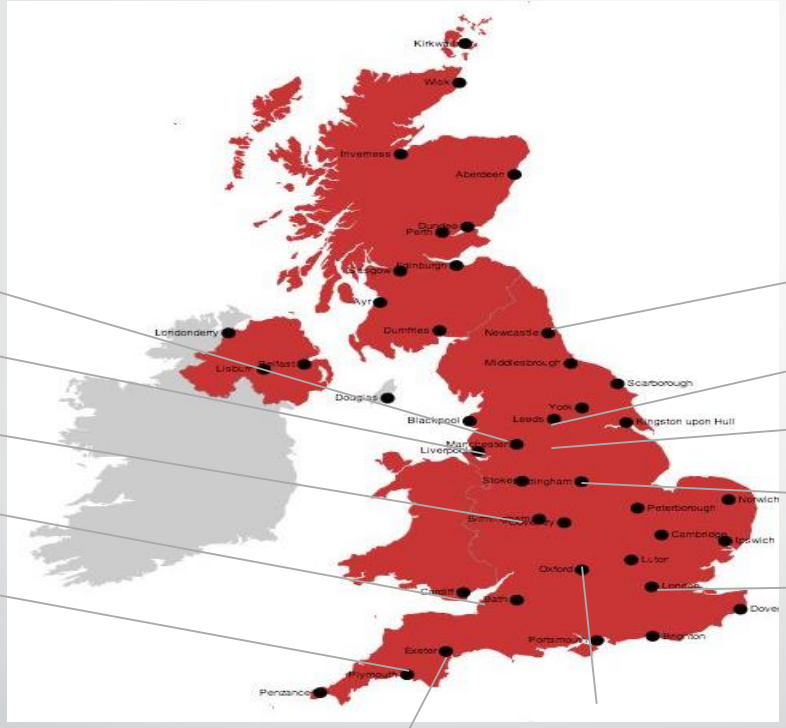
- 1-2% de tous les cancers (0,2% pour les sarcomes osseux)
- Tumeur maligne mésenchymateuse (origine tissus conjonctif)
- Incidence tumeur bénigne: 3/1000/an
- Incidence tumeur maligne: 2/100'000/an
- Donc 2 masses sur 300 est un sarcome!

Bone sarcoma

1 center per 8.8’000’000

STS

1 center per 3.8’000’000



Manchester

Liverpool (STS)

Birmingham

Bristol (STS)

Plymouth (STS)

Exeter (STS)

Oxford

Newcastle

Leeds (STS)

Sheffield (STS)

Nottingham (STS)

London

1 center / 1’000’000

Uni Basel

Uni Zurich

Uni Bern

St-Gallen

Uni Genève

Winterthur

Chur

Uni Lausanne

78. Jahreskongress | Congrès annuel – swiss orthopaedics

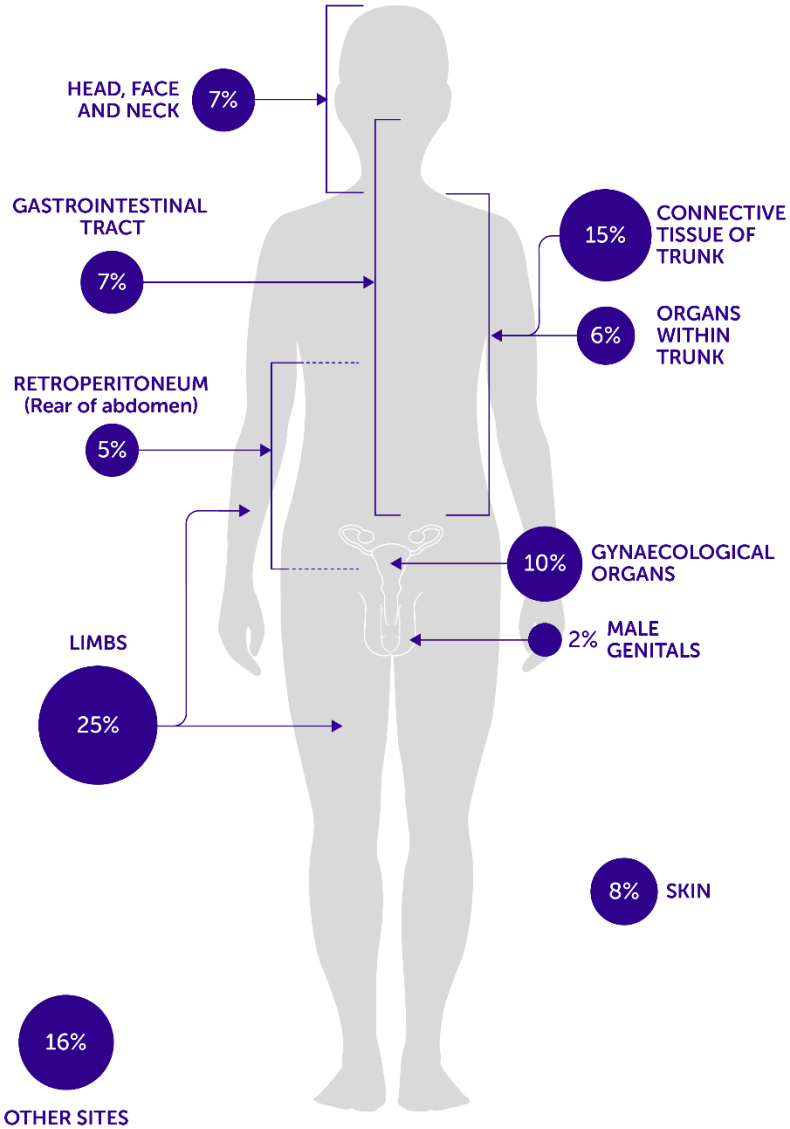
- **Lausanne 2017**

- **74 new cases**
- 5 pediatric
- 13 bone sarcomas
- 34 extremity STS

CHUV Centre hospitalier universitaire vaudois

UNIL | Université de Lausanne

SOFT TISSUE SARCOMA CASES: PERCENTAGE DISTRIBUTION BY ANATOMICAL SITE



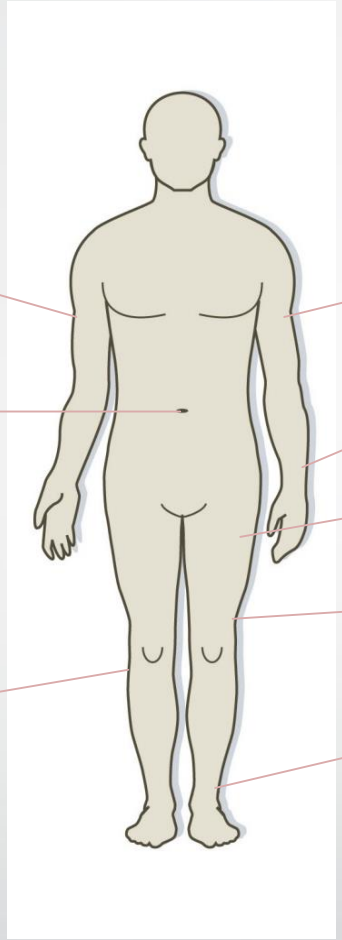
LET'S BEAT CANCER SOONER
cruk.org/cancerstats

CANCER RESEARCH UK

Membre sup. – 15%

Rachis – 3%

Membre inf. – **82%**



Epaule/bras 7.5%

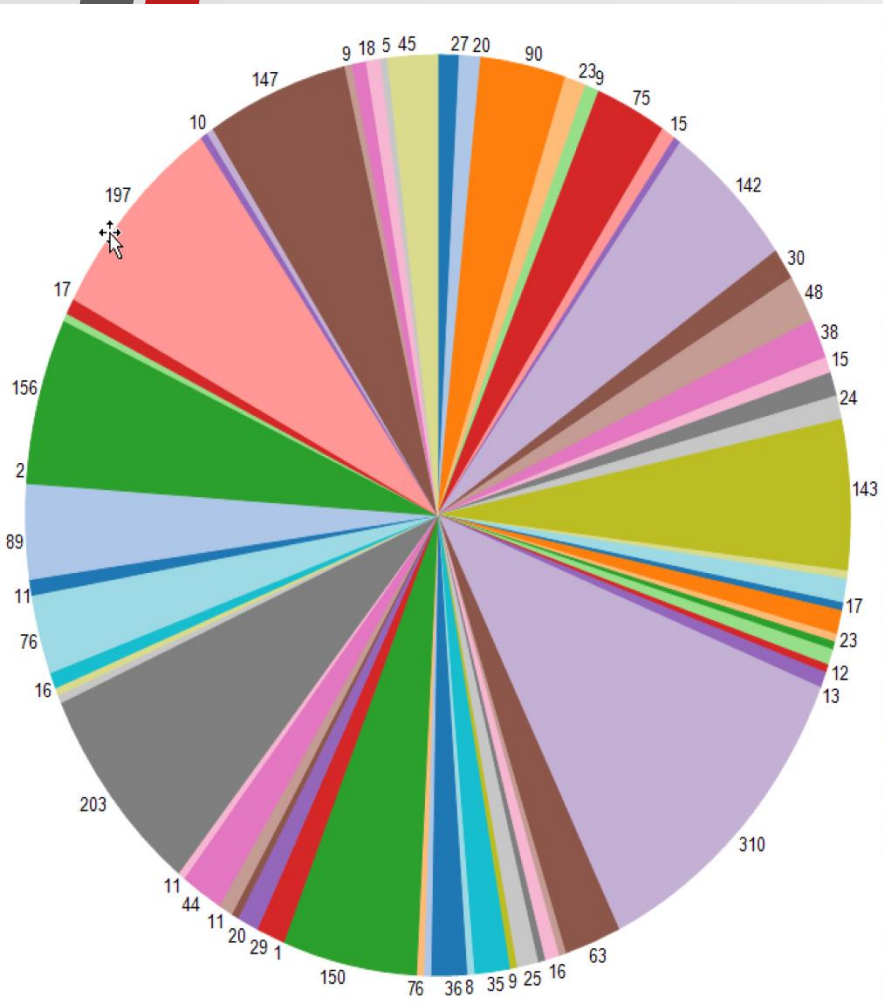
Avant-bras 7.5%

Pelvis 9%

Cuisse 52%

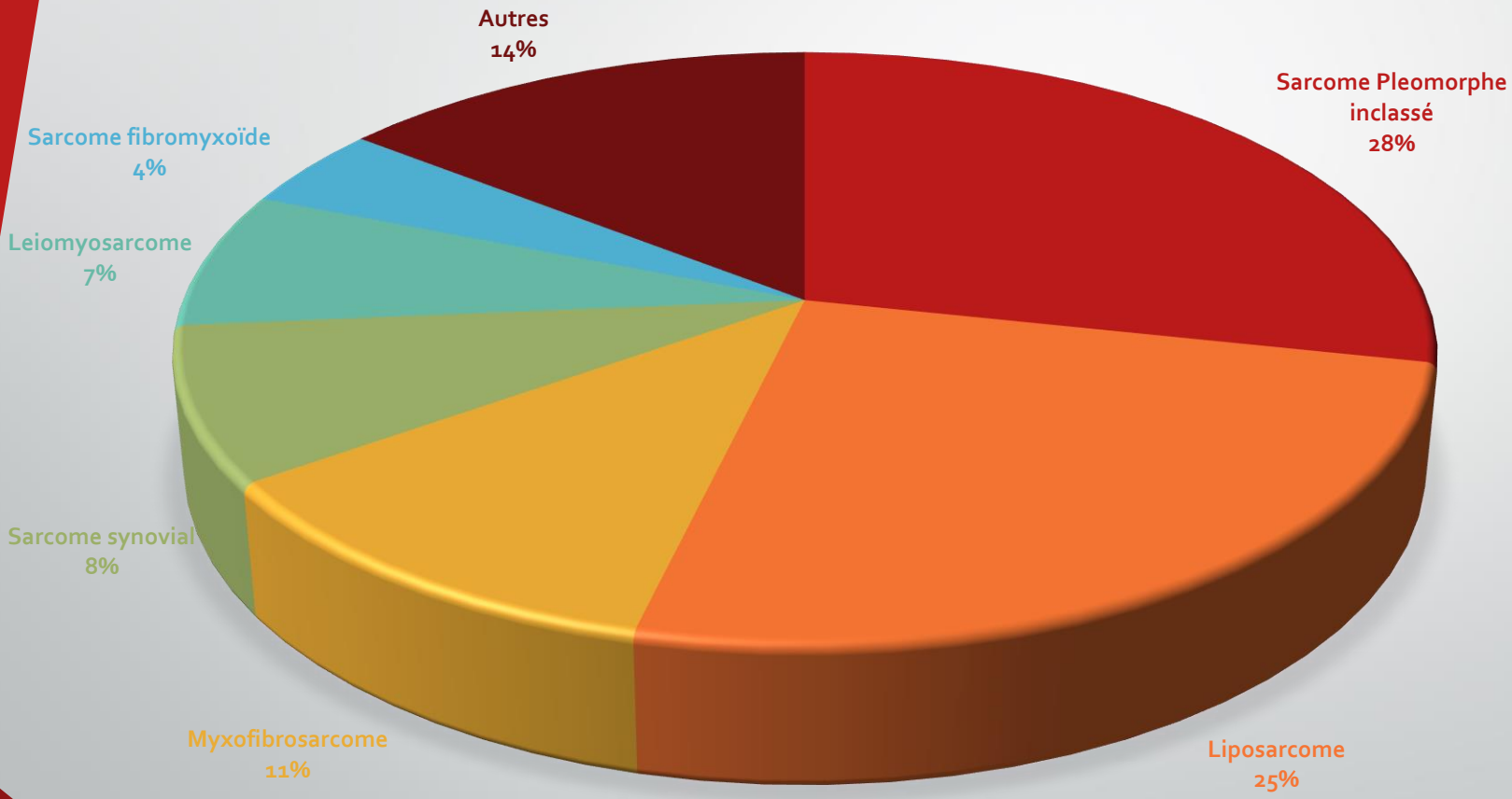
Jambe/pied 24%

- 56% hommes
- Age moyen: 54 ans (12-94)



- Alveolar Rhabdomyosarcoma
- Alveolar Soft Part Sarcoma
- Angiosarcoma
- Breast Angiosarcoma
- Breast Sarcoma
- Chondroblastic Osteosarcoma
- Chondrosarcoma
- Clear Cell Sarcoma
- Dedifferentiated Chondrosarcoma
- Dedifferentiated Liposarcoma
- Desmoid/Aggressive Fibromatosis
- Desmoplastic Small-Round-Cell Tumor
- Embryonal Rhabdomyosarcoma
- Endometrial Stromal Sarcoma
- Epithelioid Hemangioendothelioma
- Epithelioid Sarcoma
- Ewing Sarcoma
- Extraskeletal Myxoid Chondrosarcoma
- Fibroblastic Osteosarcoma
- Fibrosarcoma
- Follicular Dendritic Cell Sarcoma
- Glomangiosarcoma
- Hemangioma
- High-Grade Endometrial Stromal Sarcoma
- High-Grade Surface Osteosarcoma
- Histiocytic Dendritic Cell Sarcoma
- Inflammatory Myofibroblastic Tumor
- Interdigitating Dendritic Cell Sarcoma
- Intimal Sarcoma
- Leiomyosarcoma
- Liposarcoma
- Liver Angiosarcoma
- Low-Grade Central Osteosarcoma
- Low-Grade Endometrial Stromal Sarcoma
- Low-Grade Fibromyxoid Sarcoma
- Malignant Phyllodes Tumor of the Breast
- Mesenchymal Chondrosarcoma
- Myofibroma
- Myxofibrosarcoma
- Myxoid Chondrosarcoma
- Myxoid/Round-Cell Liposarcoma
- Myxoma
- Ossifying Fibromyxoid Tumor
- Osteoblastic Osteosarcoma
- Osteosarcoma
- Parosteal Osteosarcoma
- Perivascular Epithelioid Cell Tumor
- Phyllodes Tumor of the Breast
- Pleomorphic Liposarcoma
- Pleomorphic Rhabdomyosarcoma
- Proximal-Type Epithelioid Sarcoma
- Radiation-Associated Sarcoma
- Rhabdomyosarcoma
- Round Cell Sarcoma, NOS
- Sarcoma, NOS
- Sclerosing Epithelioid Fibrosarcoma
- Secondary Osteosarcoma
- Small Cell Osteosarcoma
- Soft Tissue Myoepithelial Carcinoma
- Solitary Fibrous Tumor/Hemangiopericytoma
- Spindle Cell Rhabdomyosarcoma
- Synovial Sarcoma
- Telangiectatic Osteosarcoma
- Tenosynovial Giant Cell Tumor Diffuse Type
- Undifferentiated Pleomorphic Sarcoma/MFH/HGSCS
- Undifferentiated Uterine Sarcoma
- Uterine Adenosarcoma
- Uterine Carcinosarcoma/Uterine Malignant Mixed Mullerian Tumor
- Uterine Epithelioid Leiomyosarcoma
- Uterine Leiomyoma
- Uterine Leiomyosarcoma
- Uterine Myxoid Leiomyosarcoma
- Uterine Perivascular Epithelioid Cell Tumor
- Uterine Sarcoma, Other
- Uterine Sarcoma/Mesenchymal
- Uterine Smooth Muscle Tumor
- Uterine Smooth Muscle Tumor of Uncertain Malignant Potential
- Well-Differentiated Liposarcoma

TYPES STS LAUSANNE 2000-2016

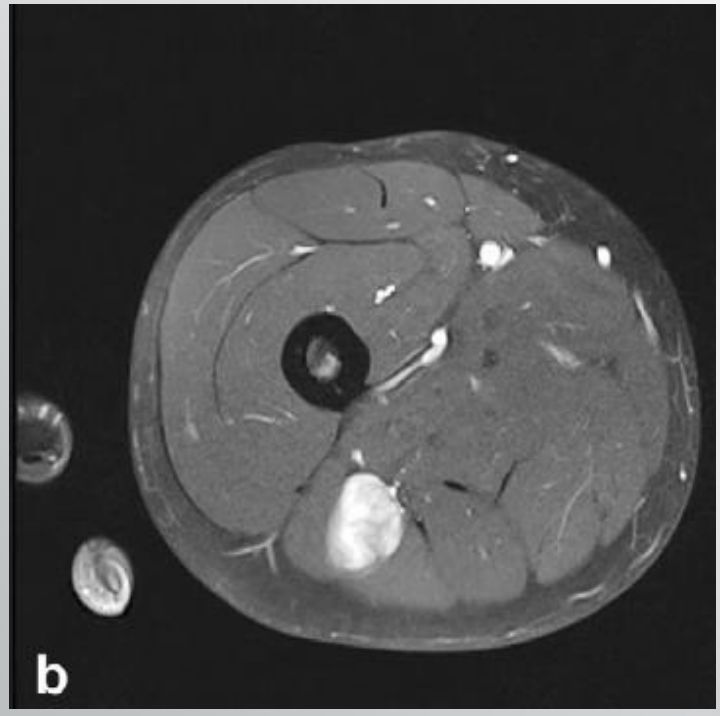


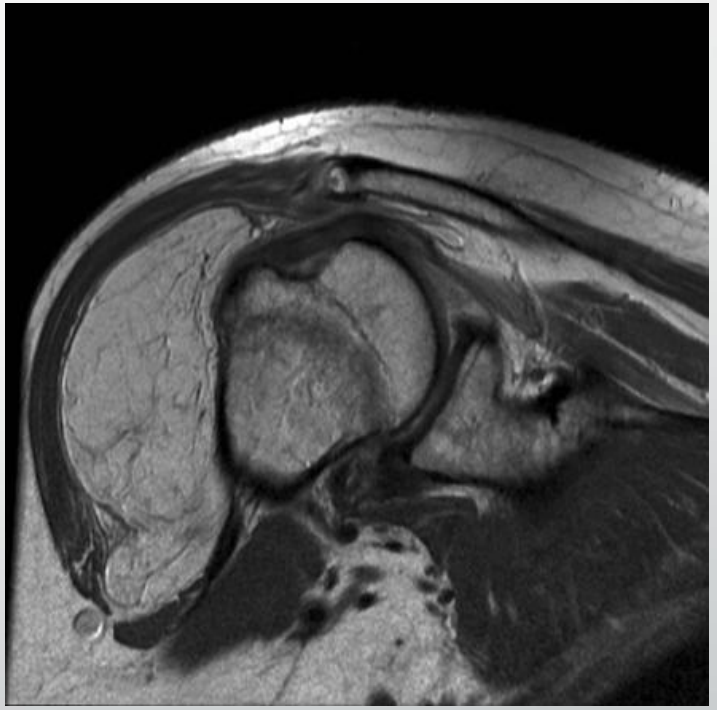
- Degré de malignité:
  - - Bénin (lipome, fibrome, angiome, ….)
  - - Intermédiaire
    - - Localement agressif: Tumeur desmoïde, Tumeur à cellule géante
    - - Faible risque métastatique: WDLPS/ATL, chondrosarcome gr1
  - - Malin (Sarcome pléomorphe inclassé, liposarcome gr2-3, …)
- Grade 1 (24.4%) 2 (39.1%) 3 (37.5%)
- Stade T(N)M

Présentation

- VARIABLE!!!!
  - - Découverte fortuite lors d'un examen pour autre chose
  - - Masse palpable
  - - Syndrome de compression
  - - Déficit fonctionnel
  - - Epanchement articulaire
  - - Symptômes B (perte de poids, vomissements, …)
  - - Fracture pathologique
  - - Hémoptysies
  - …

Comment différencier une tumeur bénigne d'un sarcome?





Nothing looks more similar to a lipoma than a liposarcoma!

- Every mass
  - >3-5cm
  - Symptomatic
  - Increasing in size
  - Deep to the fascia
  - Recurrent after initial excision…

… is a sarcoma until proven otherwise and must be referred to a sarcoma centre

J. Pike & al. : **Soft tissue sarcomas of the extremities : How to stay out of trouble.** *BCMJ*, Vol. 50, No. 6, July, August 2008, page(s) 310-317 Articles

If your lump is bigger than a golf ball and growing, think Sarcoma

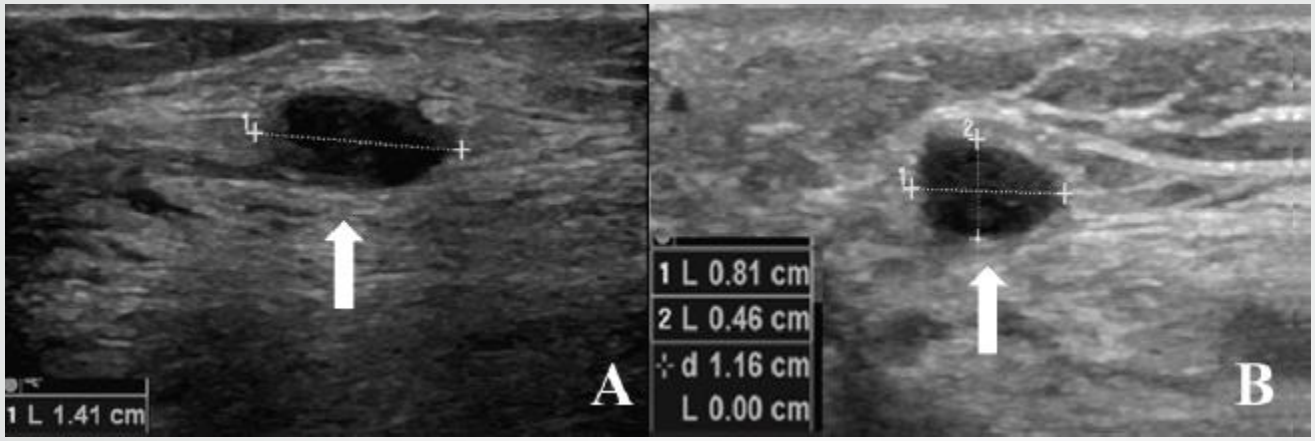
Nandra & al. : Eur. J. of Surgical oncology 2015 Oct;41(10):1400-5.
doi: 10.1016/j.ejso.2015.05.017. Epub 2015 Jun 18.

Examens complémentaires

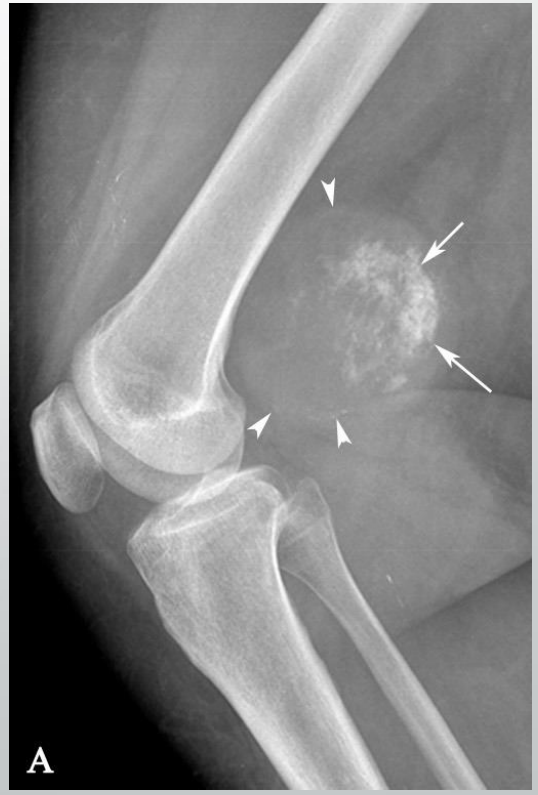
Seul l'IRM permet d'orienter vers un diagnostic, mais ne permet pas de poser le diagnostic définitif (Et ce malgré le rapport du radiologue!)

NEVER TRUST A RADIOLOGIST!

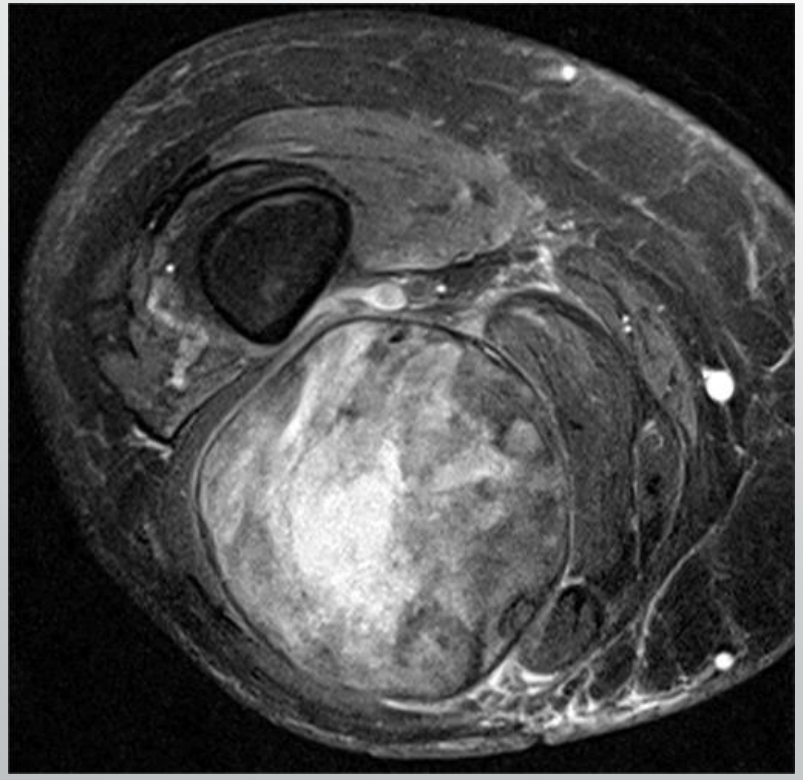
Seule la biopsie permet de poser le diagnostic



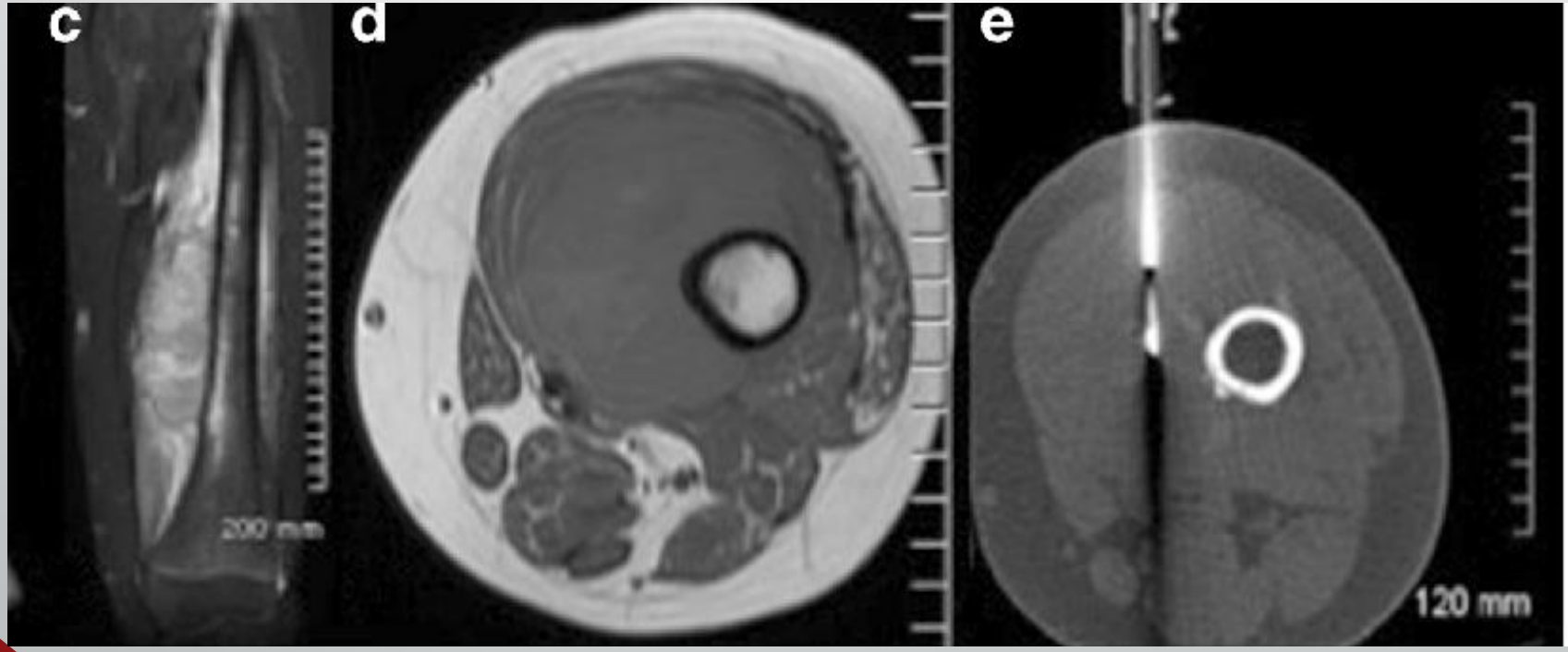
- Rx → Calcifications, voussure dans les tissus mous, …
- US → Inutile
- CT → Peu utile pour le diagnostic



- IRM:
  - - Localisation
  - - Rapport aux fascias / structures nobles
  - - Homo/hétérogénicité
  - - Taille
  - - Fenêtrage particulier (hémosidérine dans PVNS, …)



- Biopsie → Sur la voie d'abord de la future chirurgie!!!
- A l'aiguille (echo-CT guidée) ou chirurgicale
- Hémostase parfaite!
- (Bleu de méthylène)



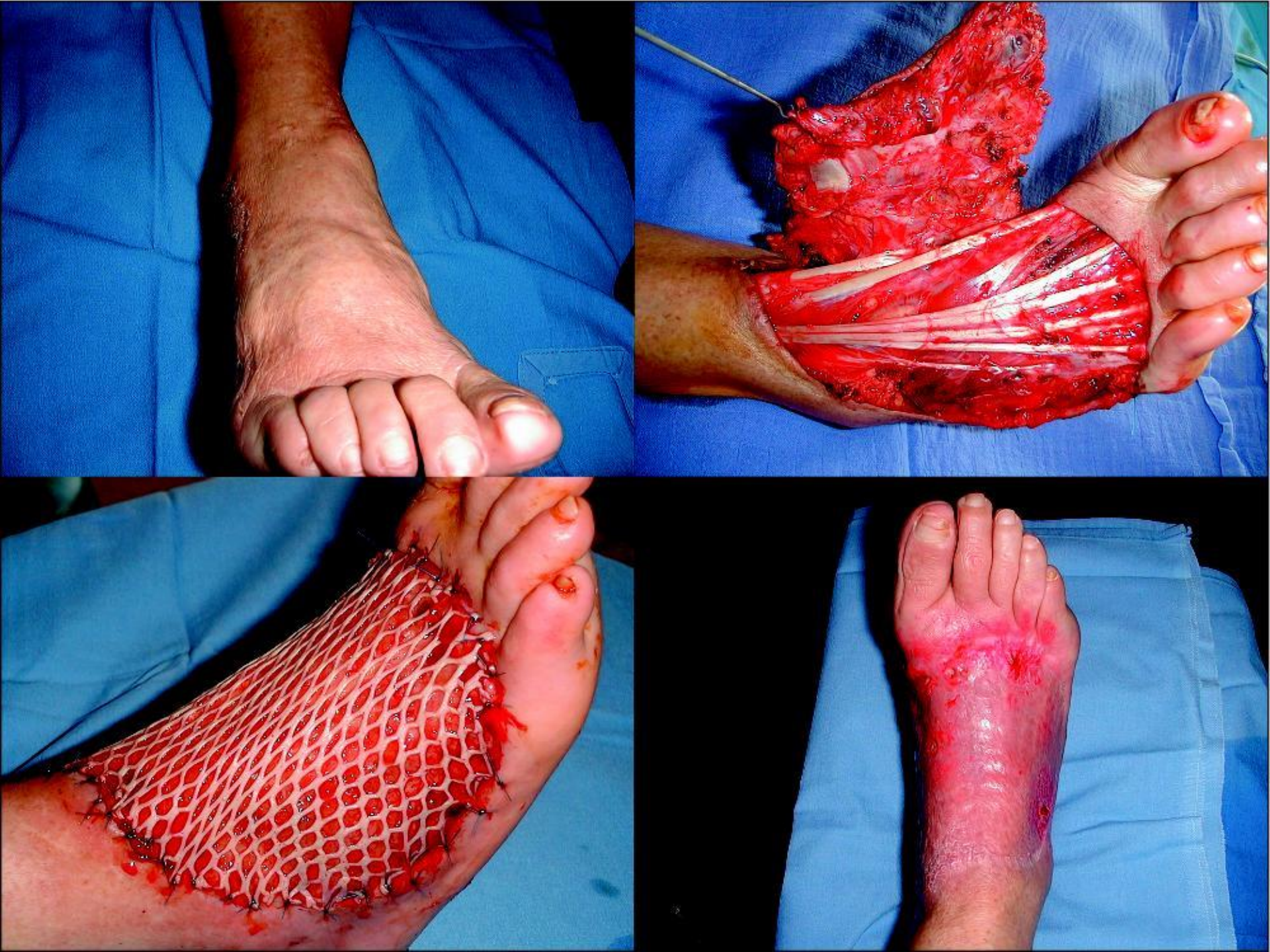
Biopsie → Pathologie: diagnostic de sarcome

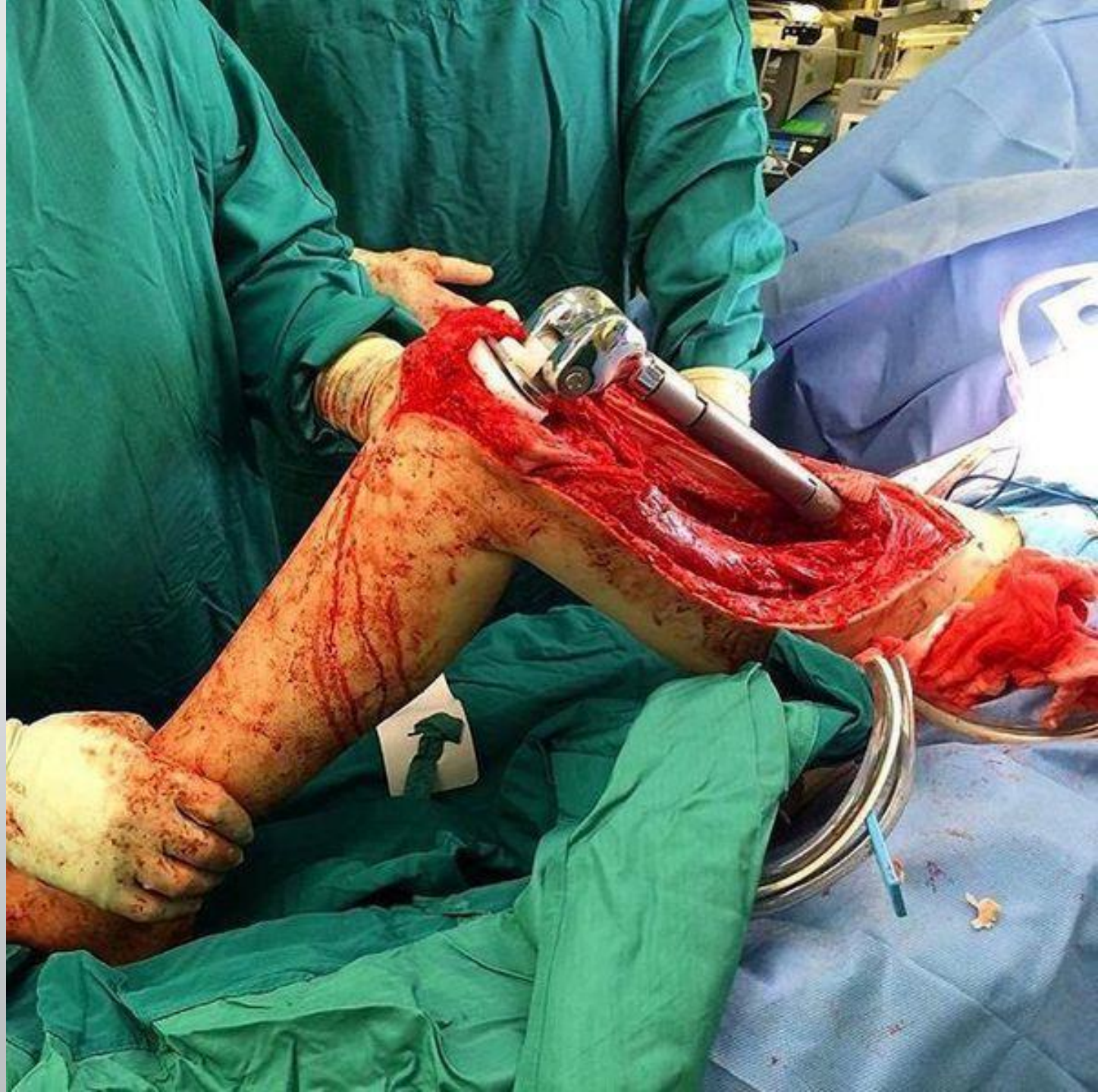
Bilan d'extension

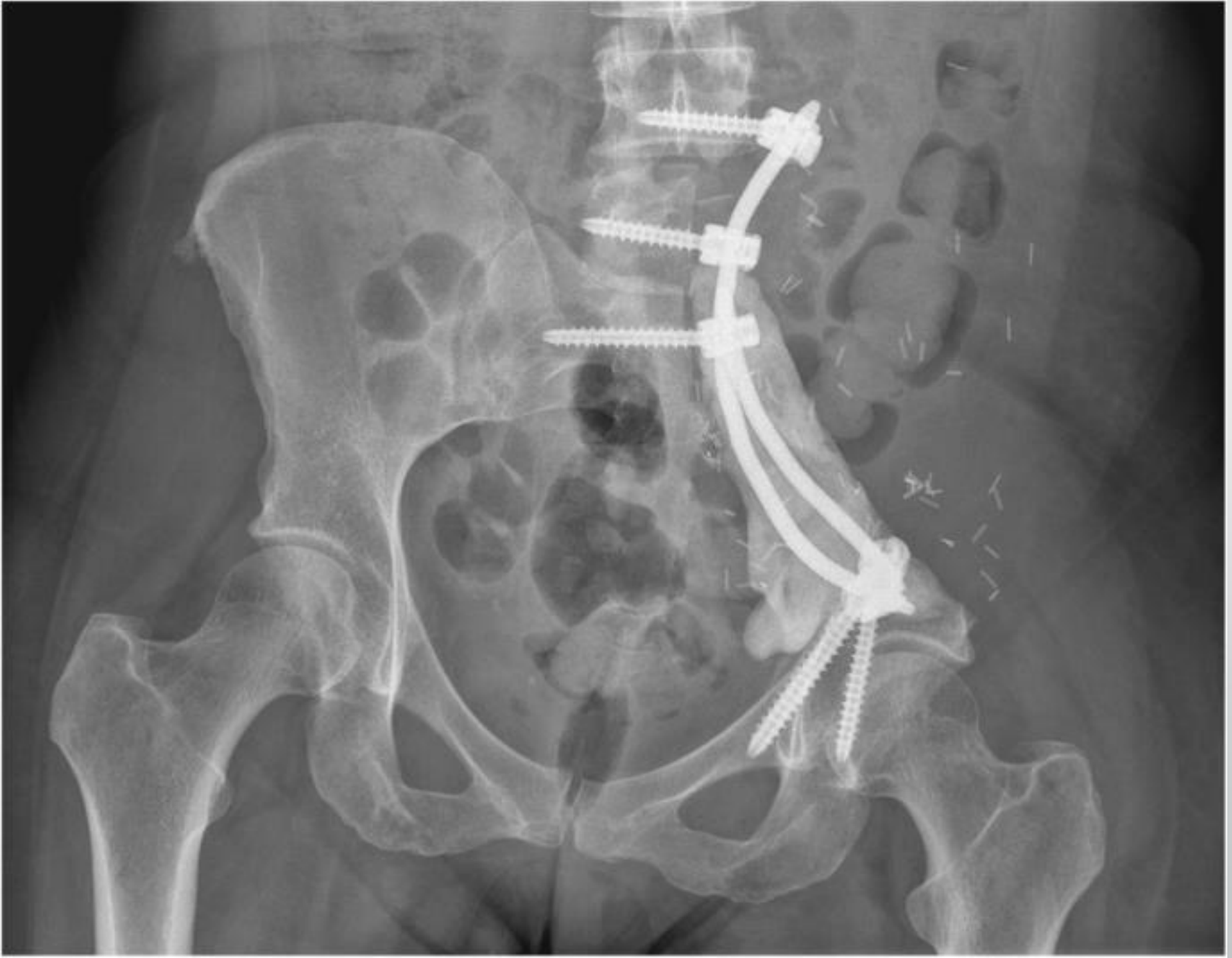
→Colloc multidisciplinaire → Plan de traitement

→Curatif? Palliatif
→Chimiothérapie? Neo ou adjuvante?
→Radiothérapie? Neo ou adjuvante?
→Autre?

→Chirurgie:
- →Excision large (R0-R1-R2)
- →Respect des fascias
- →Excision du trajet de biopsie
- →Hémostase parfaite
- →Orienter la pièce
- → Life is more important than limp







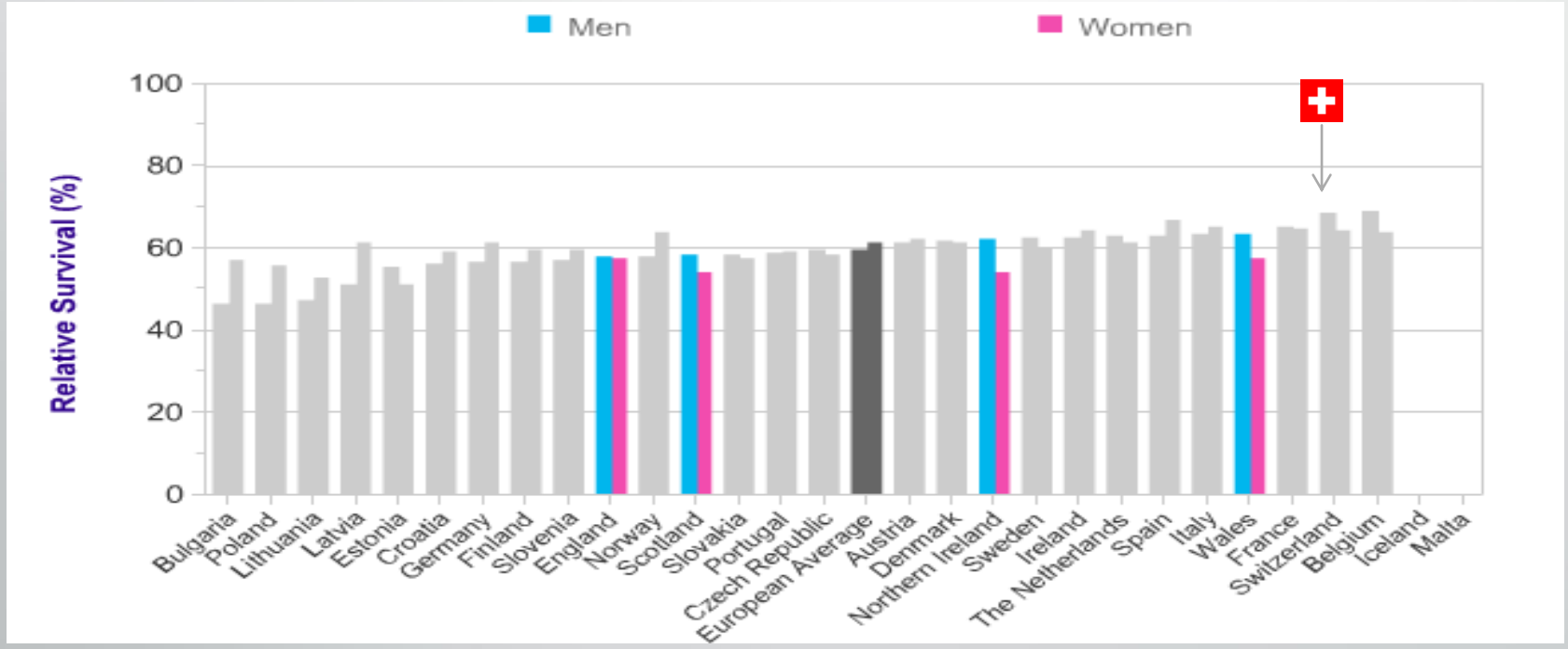
- National Cancer Institute (NCI) estimation for 2018
  - 13,040 new cases
  - 5150 deceased (39.5%)

- Canadian Cancer Society
- 5 year STS relative survival : 65%

- Rothermundt 2014
  - 174 patients
  - 21% local recurrence (31)
  - 34.7% distant recurrence (51)

Rothermund &al What is the role of routine follow-up for localised limb soft tissue sarcomas? A retrospective analysis of 174 patients; Br J Cancer 2014 May 13;110(10):2420-6

Soft tissue sarcoma, age-standardized Five-Year Relative survival, Adults (15+), European Countries, 2000-2007 (Cancer Research UK)



http://www.cancerresearchuk.org/health-professional/cancer-statistics/statistics-by-cancer-type/soft-tissue-sarcoma/survival#heading-One

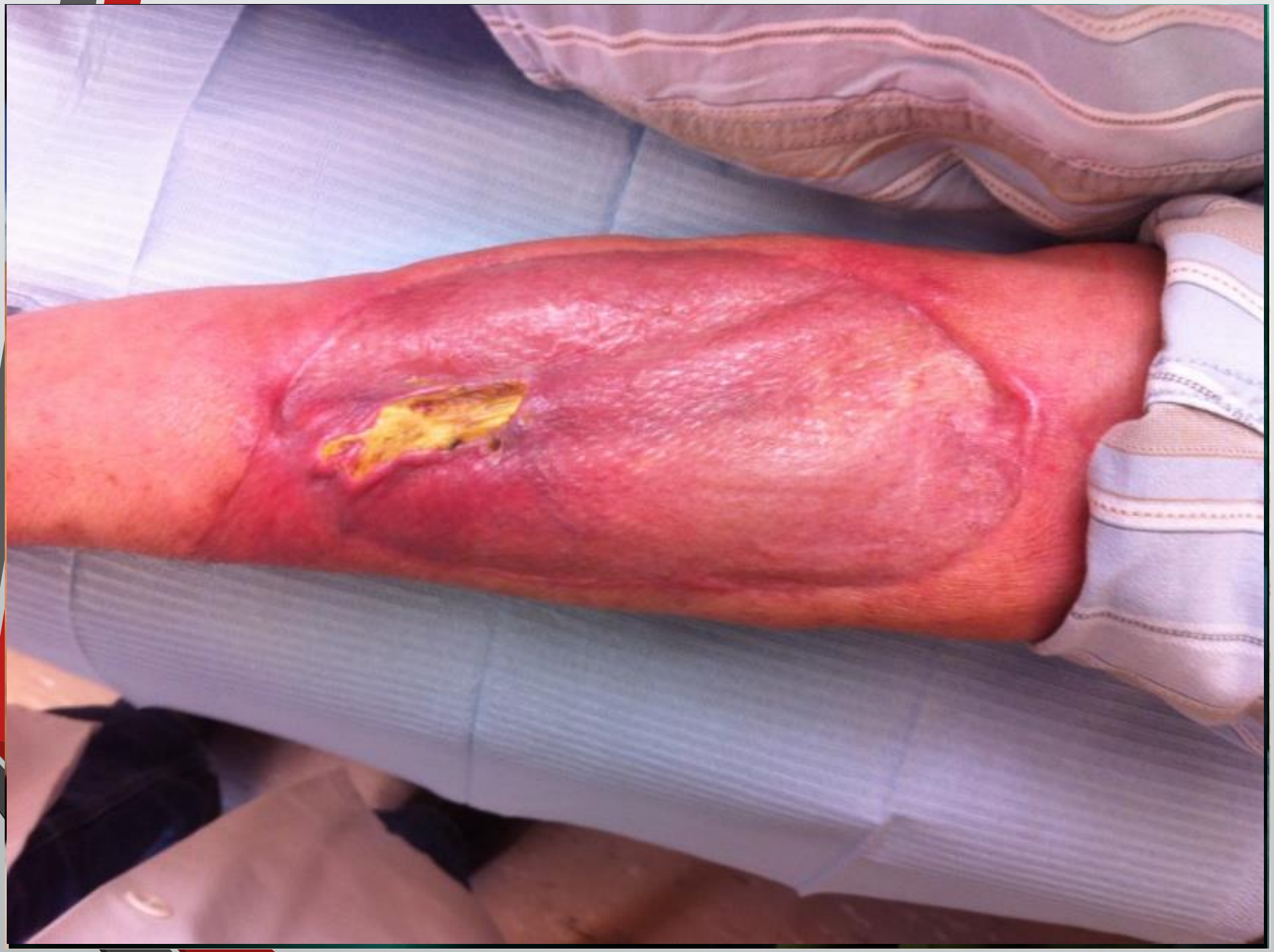
Mean Europe 59% five-year survival

OOPS!

- Excision accidentelle d'un sarcome: 31.8%!!

Bilan d'extension → Colloc multidisciplinaire → Plan de ttt

Chirurgie → Excision du lit tumoral



OOPS vs standard

- Pas de différence outcome survie, métastase, récidive
- Pas de différence outcome fonctionnel
- Moyenne 2 opérations supplémentaires
- Hospitalisation plus longue (5 jours)
- Coûts 64% plus élevés

European Journal of Surgical Oncology xxx (xxxx) xxx

Contents lists available at ScienceDirect

European Journal of Surgical Oncology

journal homepage: www.ejso.com

Oncological outcome, functional results and costs after unplanned excision of musculoskeletal soft tissue sarcoma

Boris Morattel [a, *], Laurent Mustaki [a], Michael Montemurro [b], Igor Letovanec [c], Andre D. Durham [d], Fabio Becce [e], Patrick Omoumi [e], Pietro G. di Summa [f], Maurice Matter [g], Hannes A. Rüdiger [h], Stéphane Cherix [a]

Take home message

Toute masse

- De la taille d'une **balle de golf** ou plus grosse
- Ou **sous-fasciale**
- Ou qui **grossit**
- Ou qui est **symptomatique**
- Ou qui **récidive** après une première excision

Est un sarcome jusqu'à preuve du contraire et doit être adressée dans un centre spécialisé

Seule la biopsie peut poser le diagnostic