Sarcomas of the soft tissue and bones

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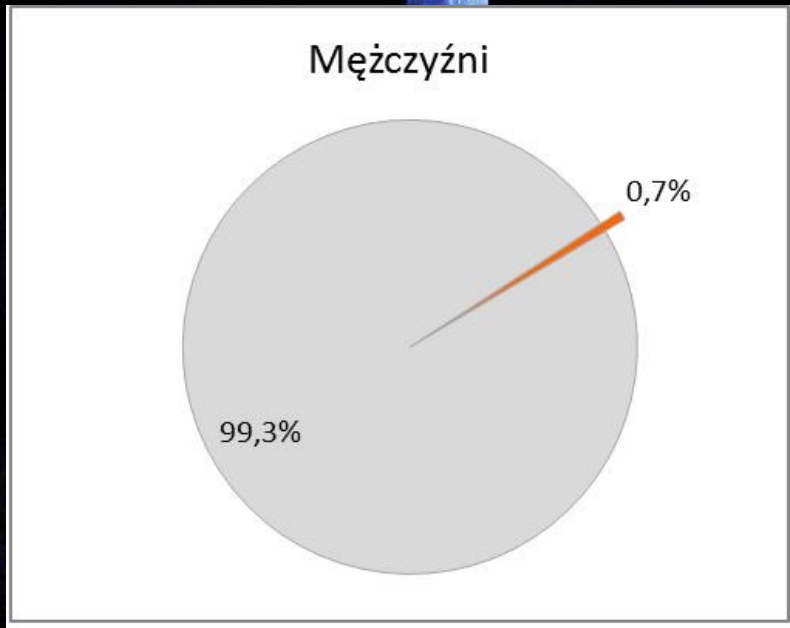
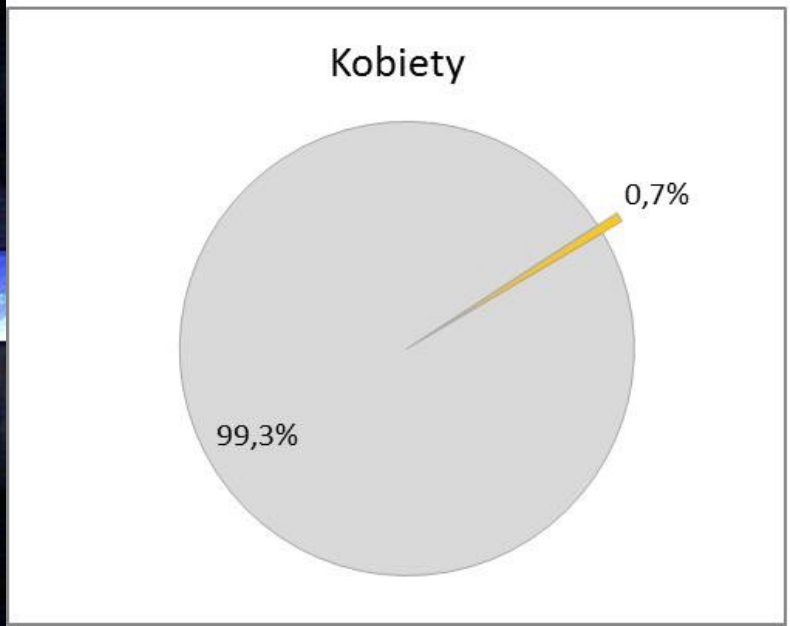
Epidemiology of malignant tumors in Poland

malignant neoplasms in Poland 2011

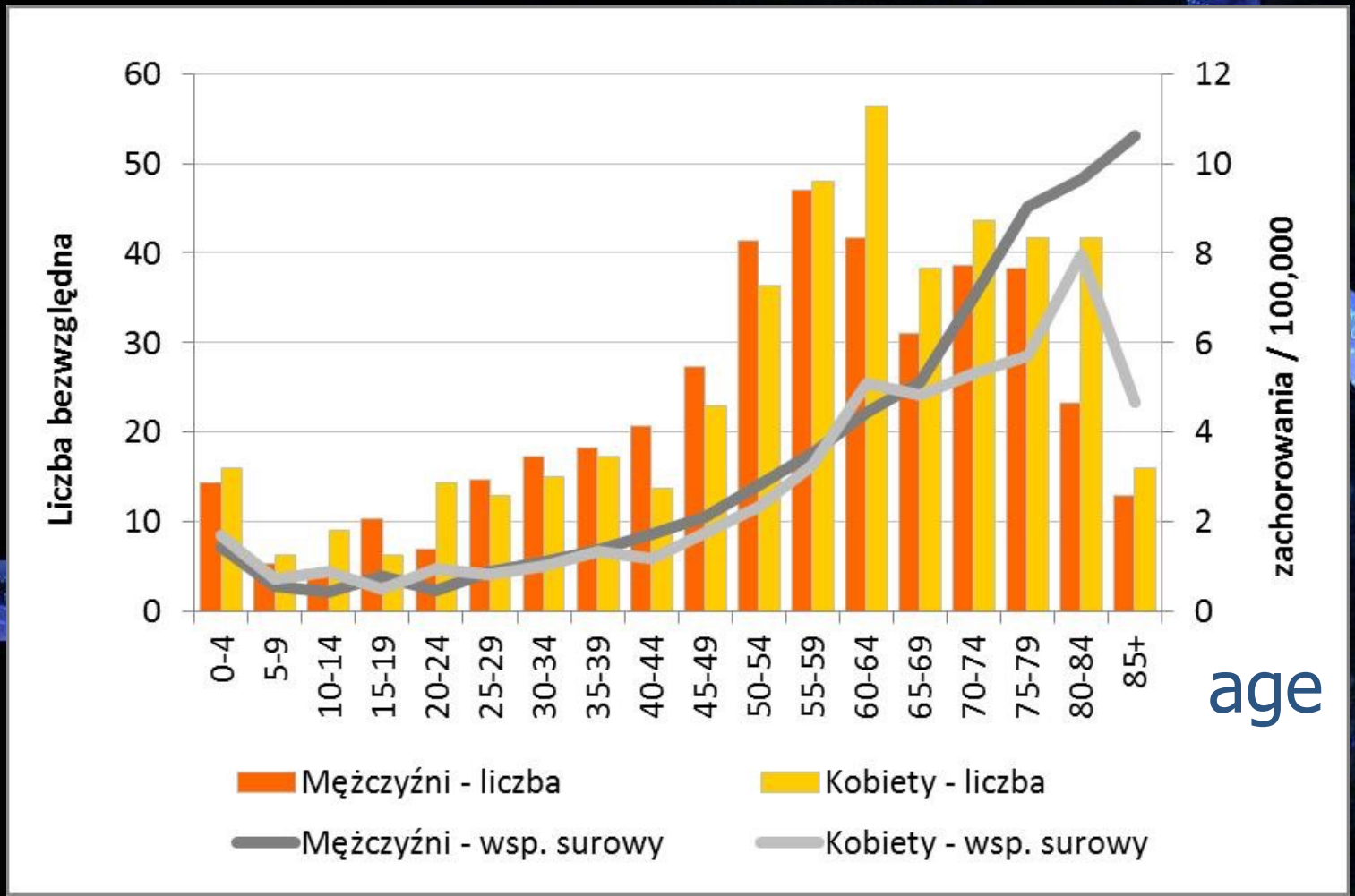
Soft tissue sarcomas (C48-C49) STS

The incidence of soft tissue cancers in Poland in 2010

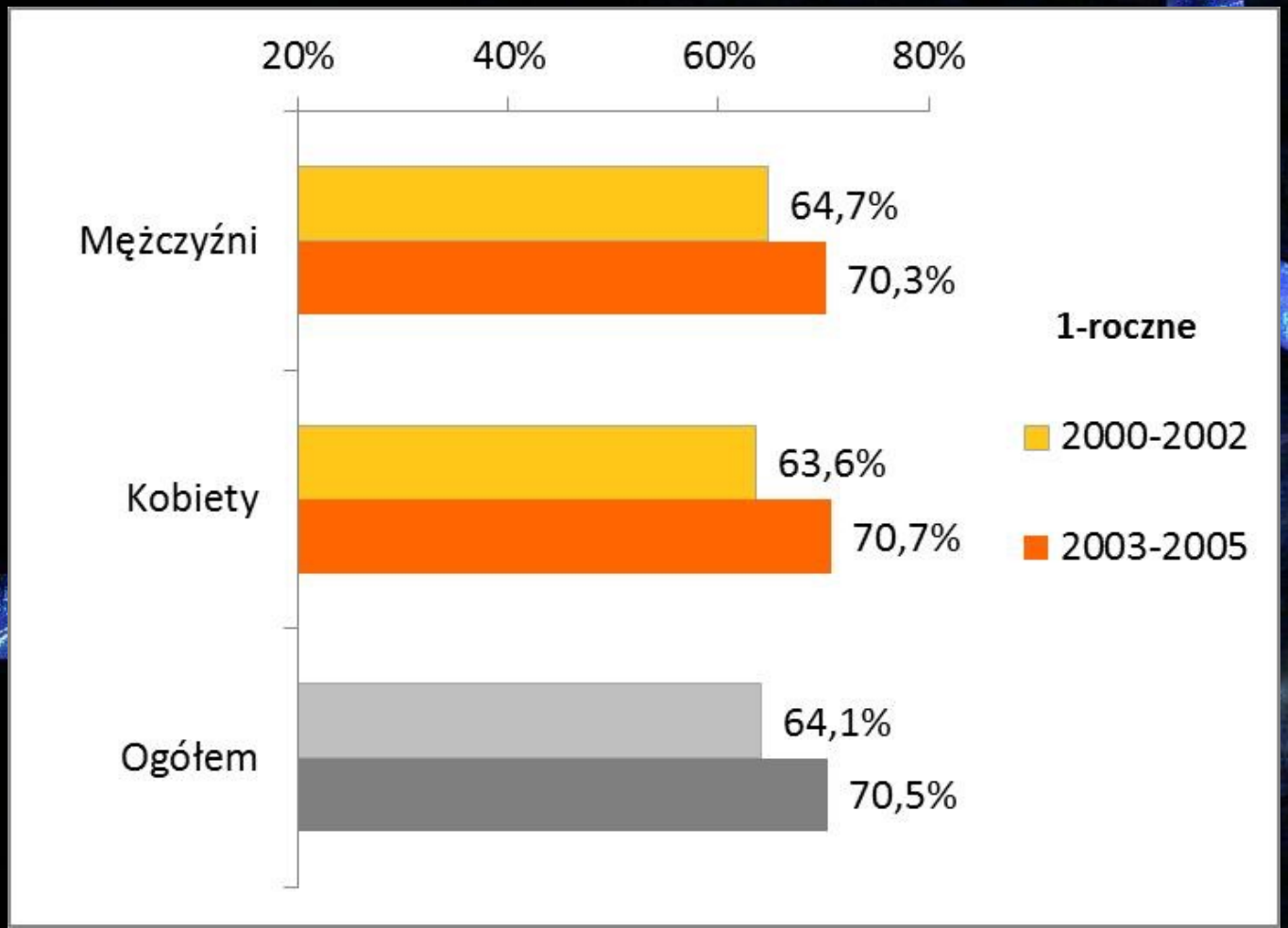




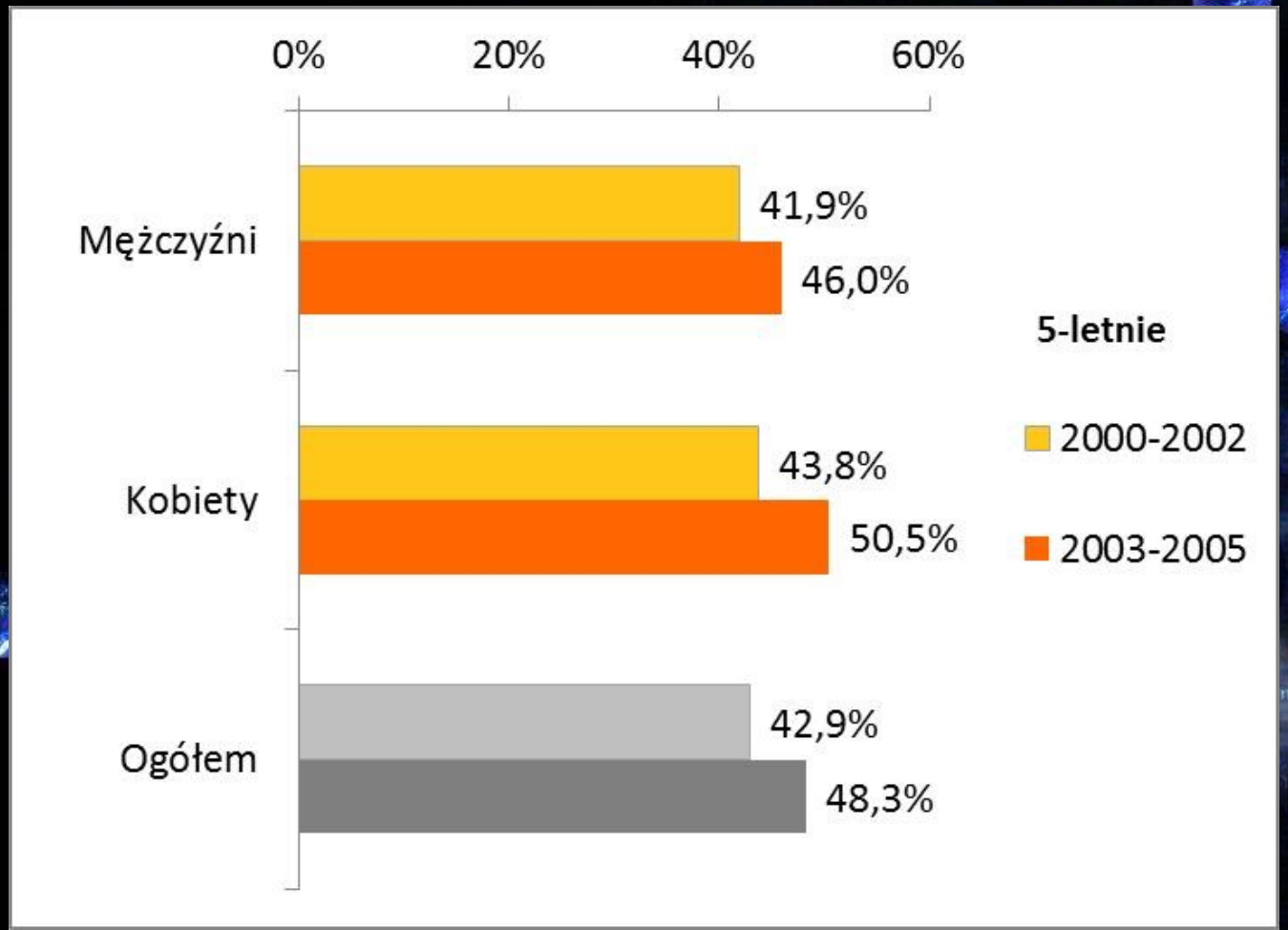
soft tissue morbidity in Poland 2008-2010



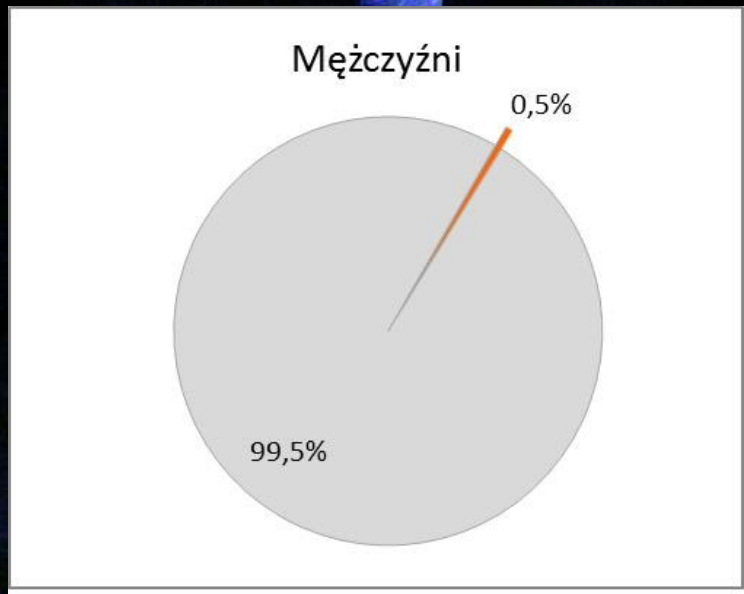
1-year survival STS (Poland)

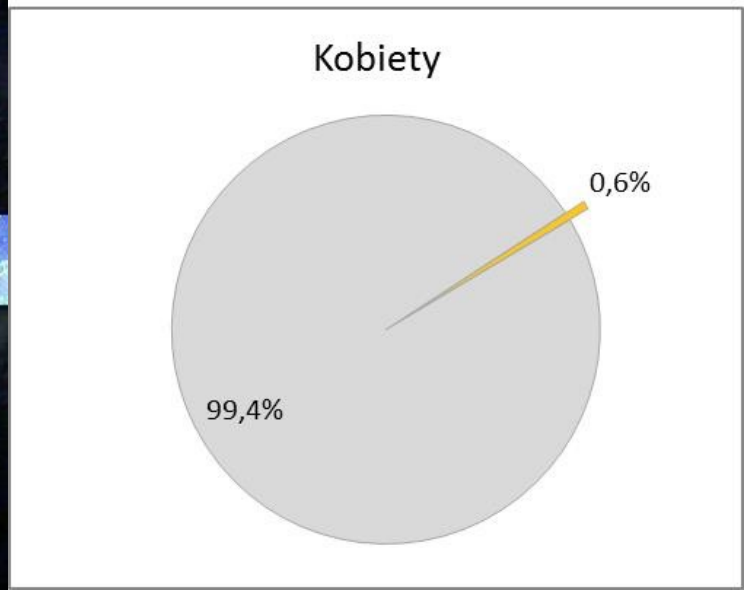


5-year survival STS (Poland)



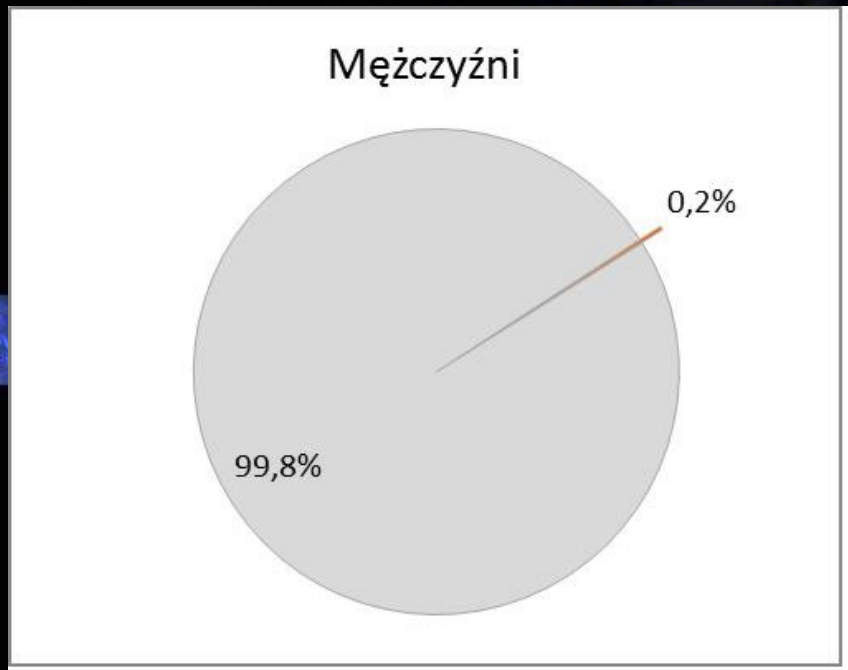
Incidence of deaths caused by STS in Poland 2010

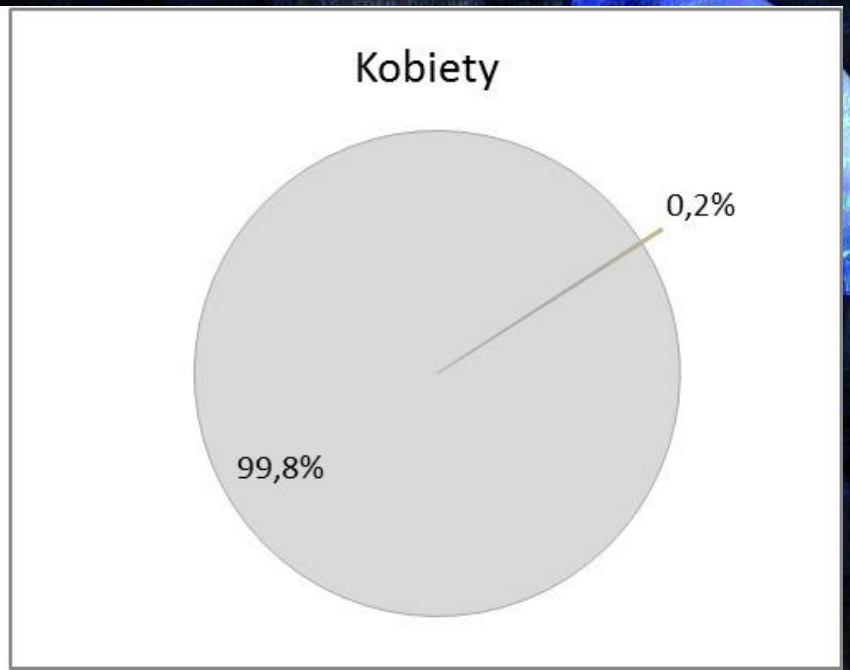




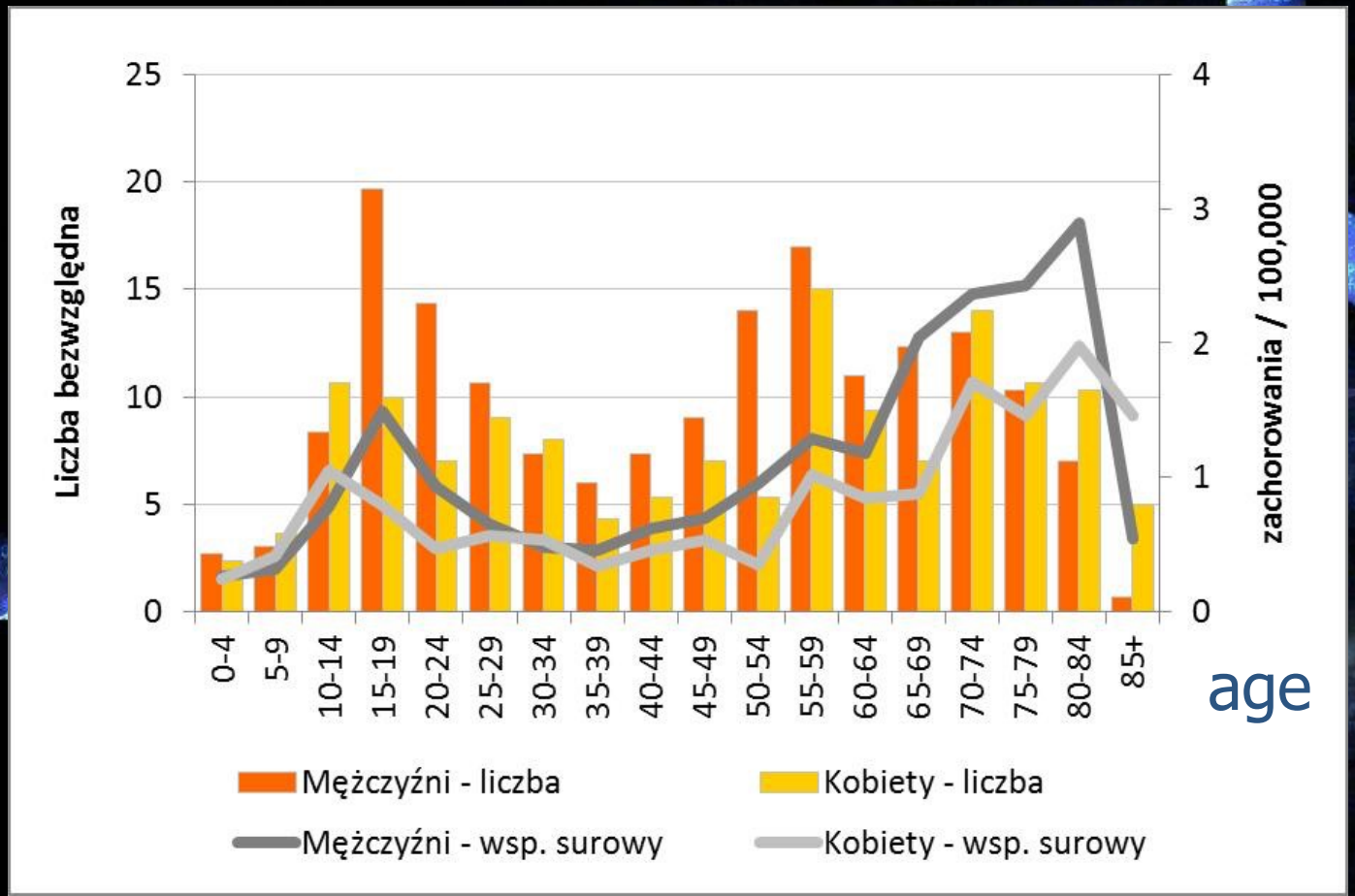
Bone sarcomas (C40-C41) BS

Incidence of morbidity - bone sarcomas

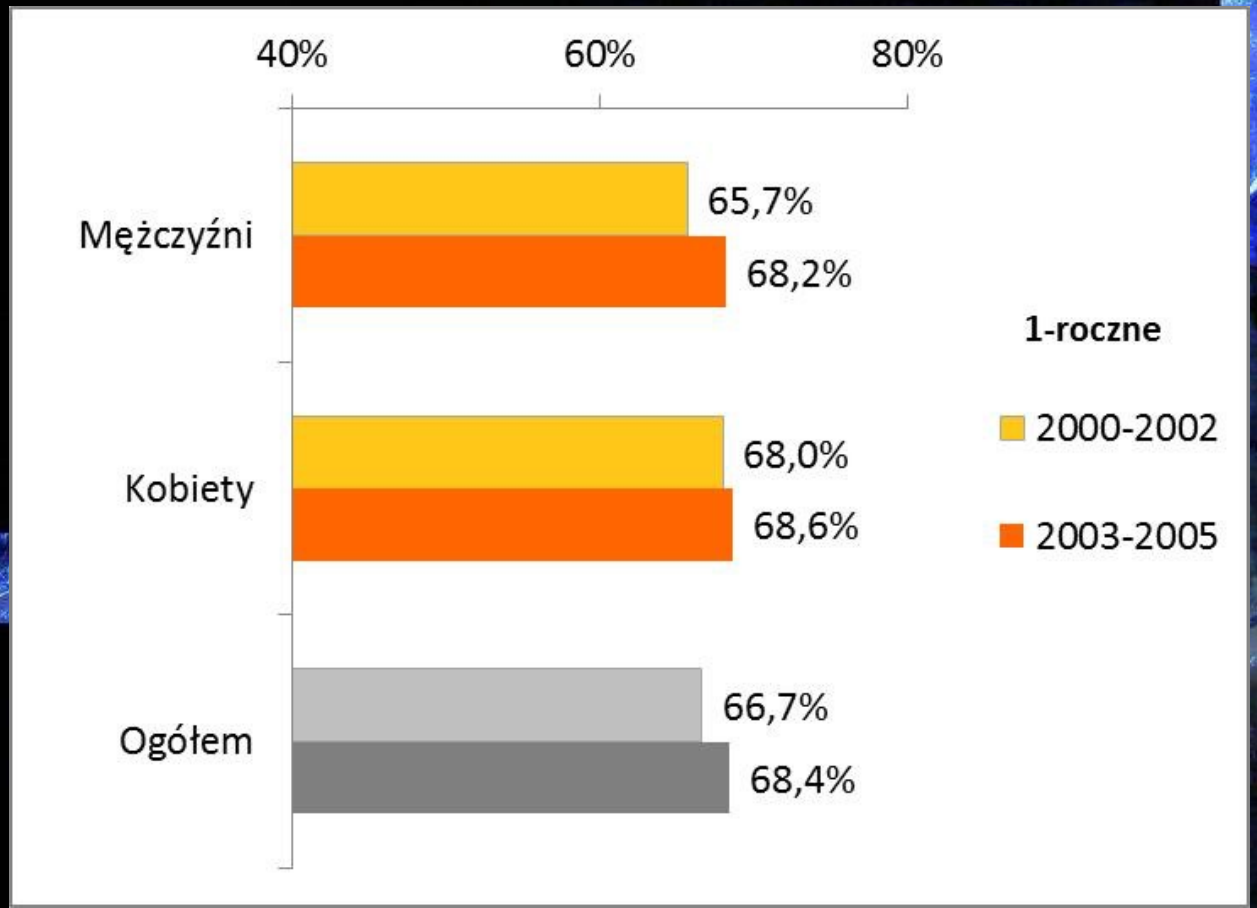




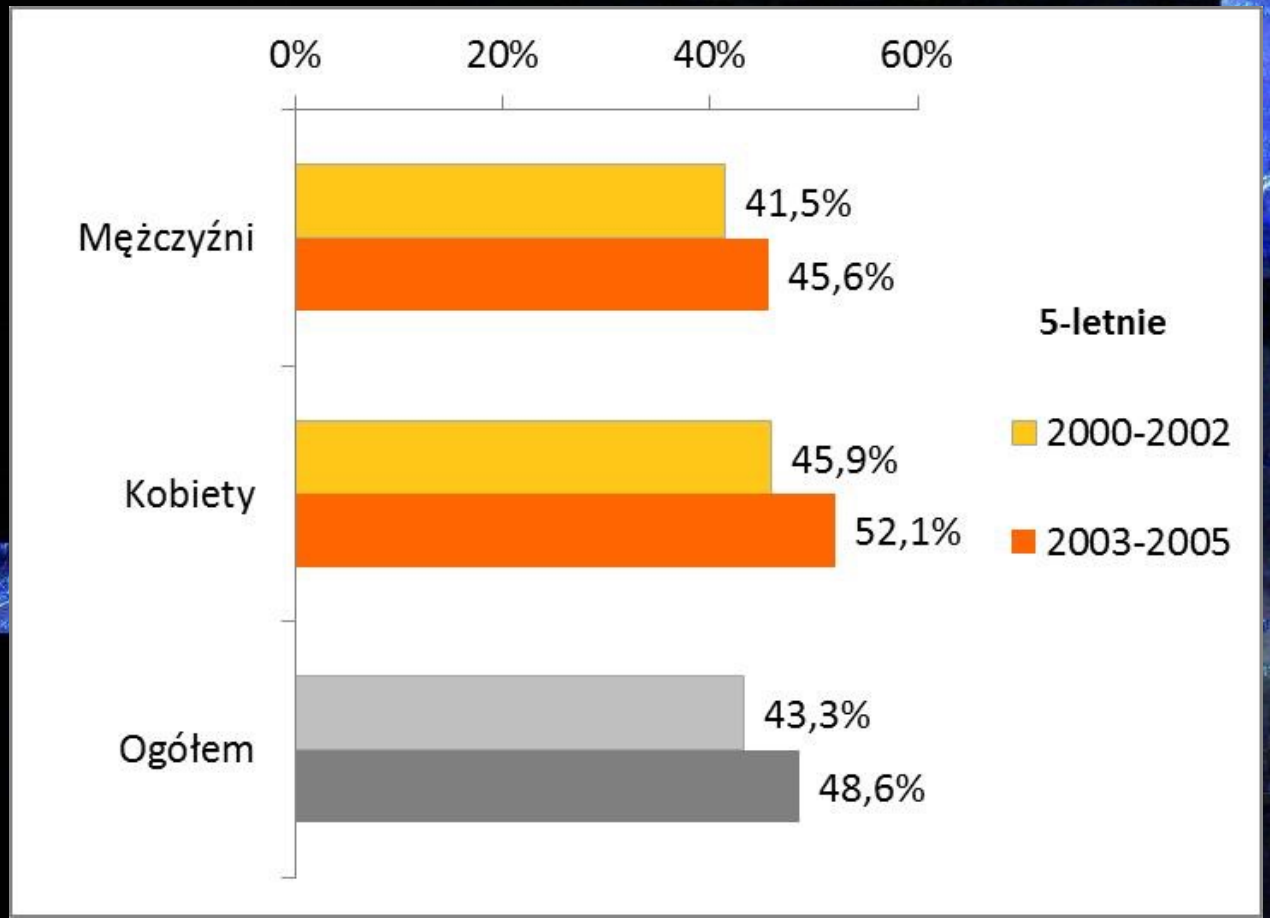
age dependent morbidity (bone and cartilage sarcoma)



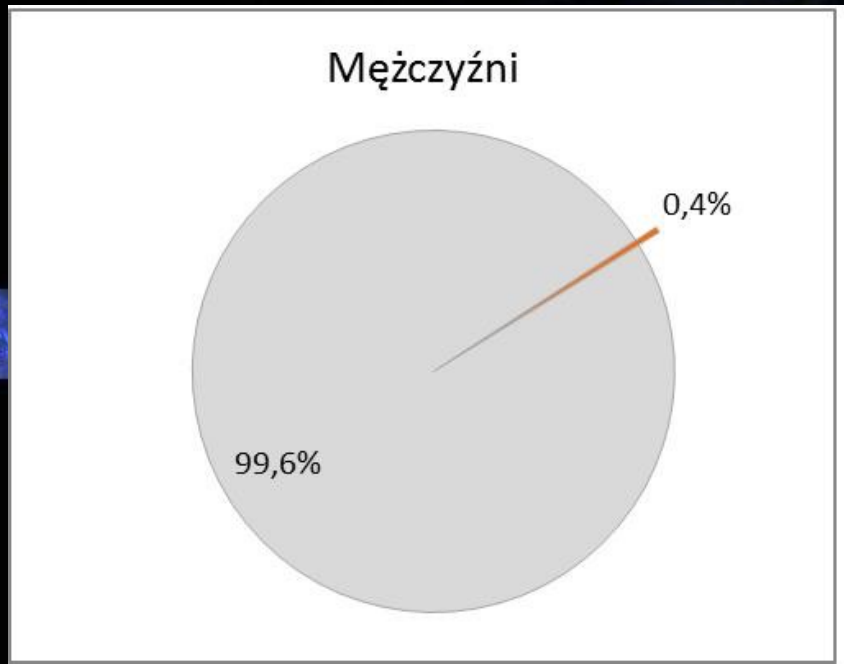
1-year survival BS (Poland)

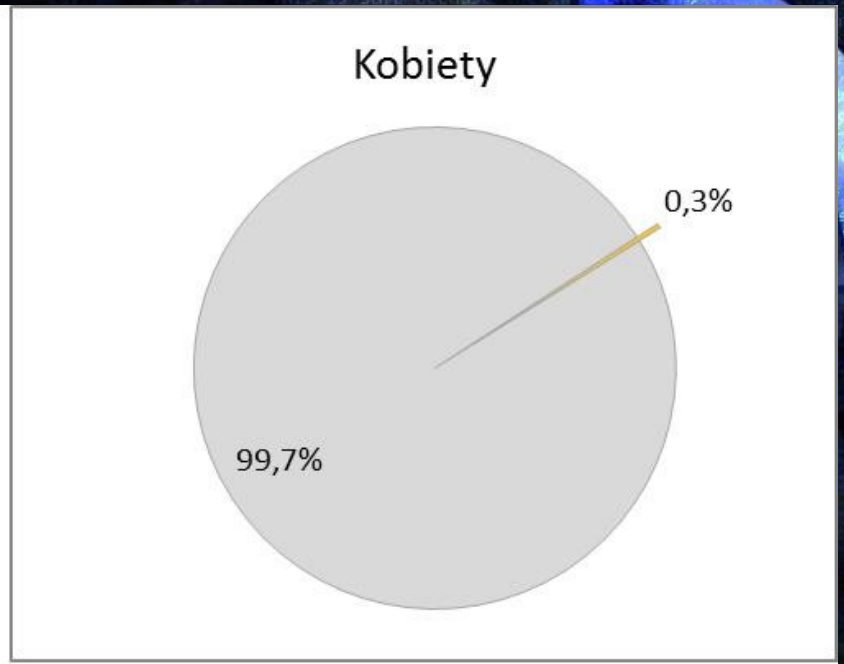


5-year survival BS (Poland)

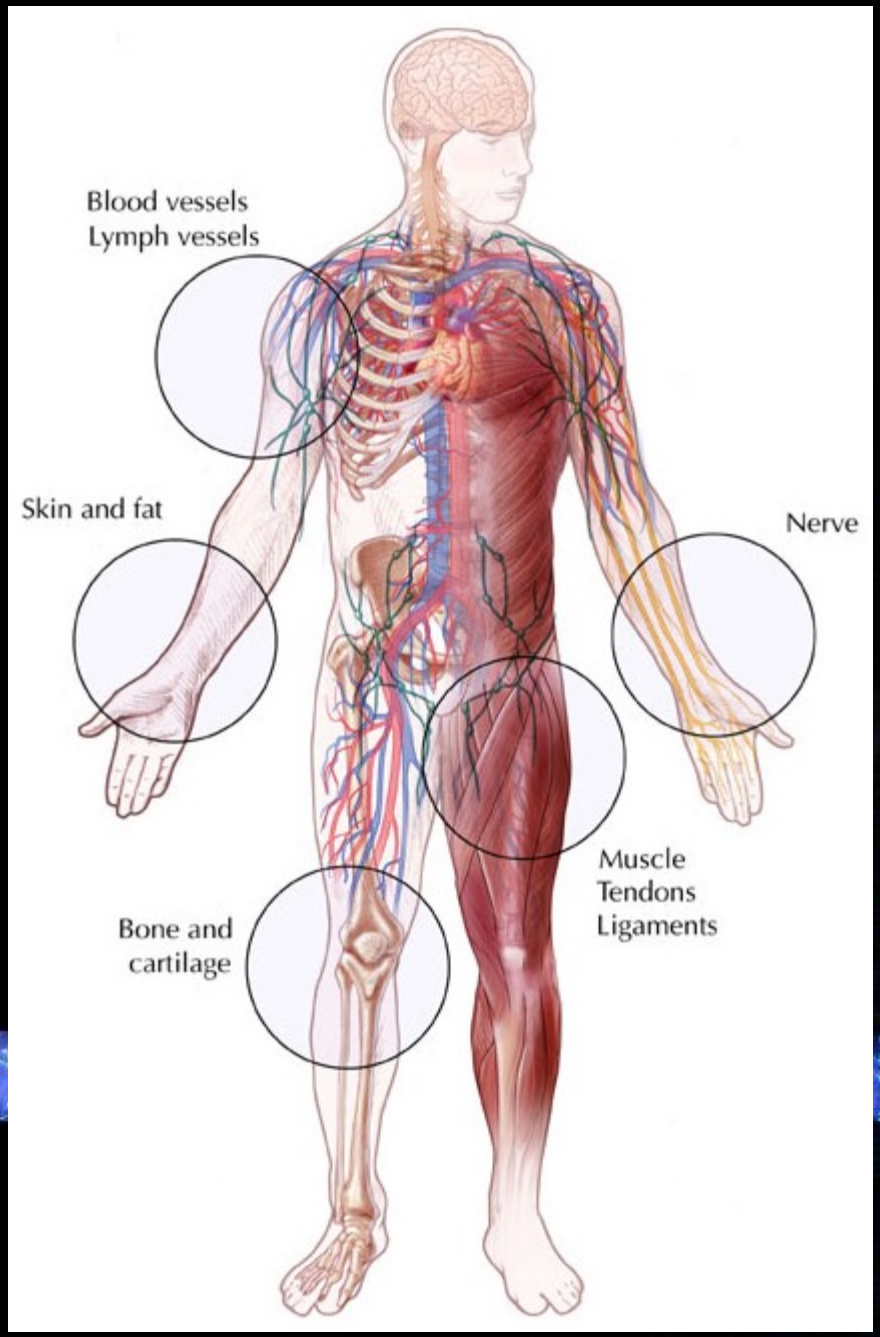


incidence of deaths caused by BS



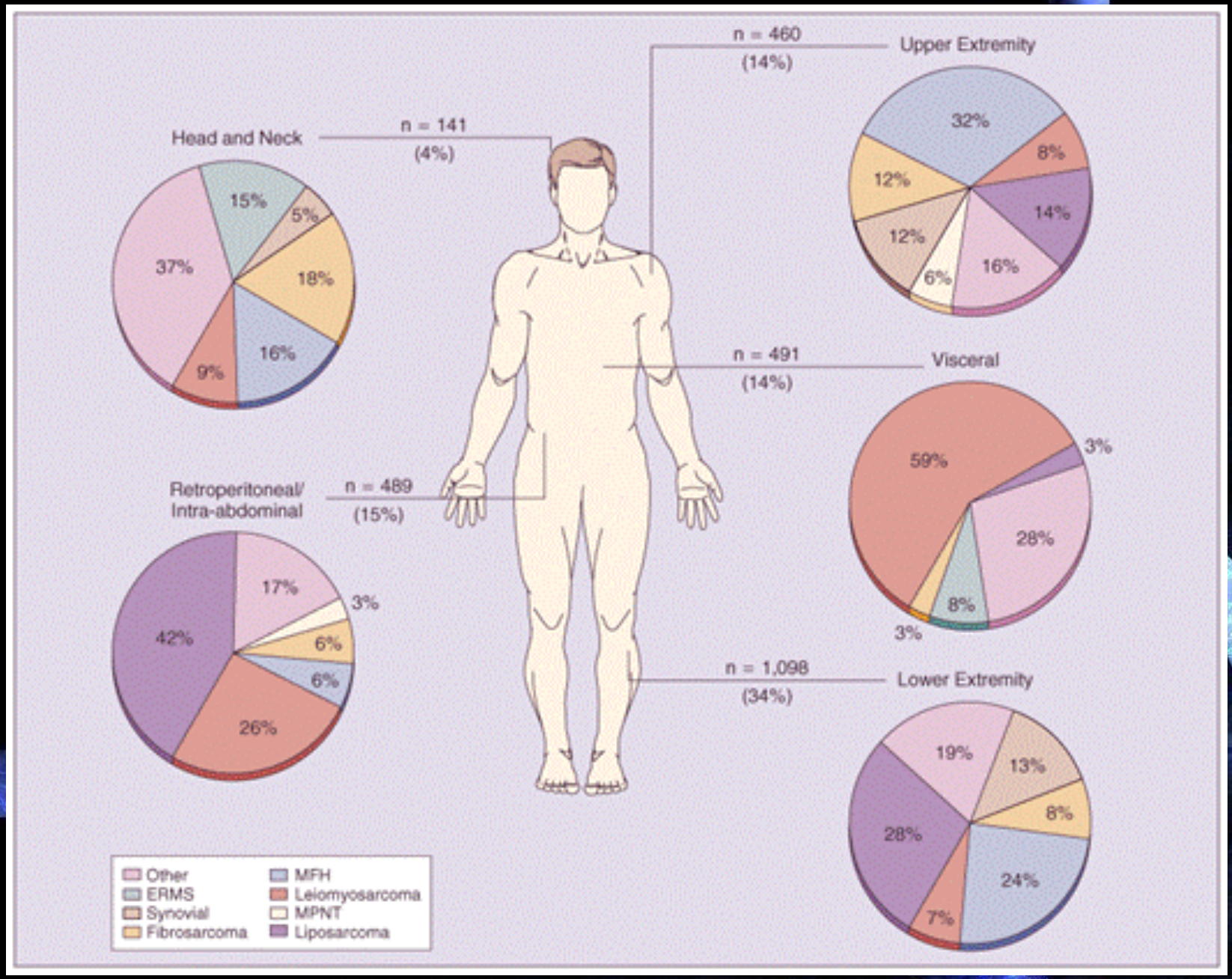


- 1 – 2% of all cancers at adult patients
- diversity in pathologic presentation:
  ca 100 subtypes
- benign soft tissues lesions are much more common than sarcomas
- there are no presarcomatous changes
- there are no in situ lesions
- early diagnosed are curable at 70%, but metastatic disease is rarely curable
- combined modality treatment:
  surgery and radiotherapy
-hematogenous spread of dissemination



distribution

- any site of the body
- 50% - in the extremities – **2/3 lower limbs**
- 30% - intraabdominal localisations


n = 460
(14%)
Upper Extremity
32%
8%
14%
16%
6%
12%
12%
n = 141
(4%)
Head and Neck
15%
5%
18%
16%
9%
37%
n = 491
(14%)
Visceral
59%
3%
28%
8%
3%
n = 489
(15%)
Retroperitoneal/
Intra-abdominal
17%
3%
6%
6%
26%
42%
n = 1,098
(34%)
Lower Extremity
19%
13%
8%
24%
7%
28%
Other
ERMS
Synovial
Fibrosarcoma
MFH
Leiomyosarcoma
MPNT
Liposarcoma

Etiology

predisposing and associated factors:
-lymphedema for lymphangiosarcoma, immunosupresion, viral infections (?)

- iatrogenic factors:
radiotherapy (breast, cervical cancers, lyphoma previously irradiated)

Etiology and genetics

genetic alterations:

- neurofibromatosis (for MPNST – Malignant Peripheral Nerve Sheath Tumors),
- Li Fraumeni syndrome,
- Gardner's syndrome (Familial Adenomatous Polyposis and desmoids)

Pathologic clasification

- According to the normal tissue the tumors mimic
- Sarcomas arrise from mesoderm
- Hematogenous spread
- Lymphatic pattern of dissemination is uncommon – 10% cases

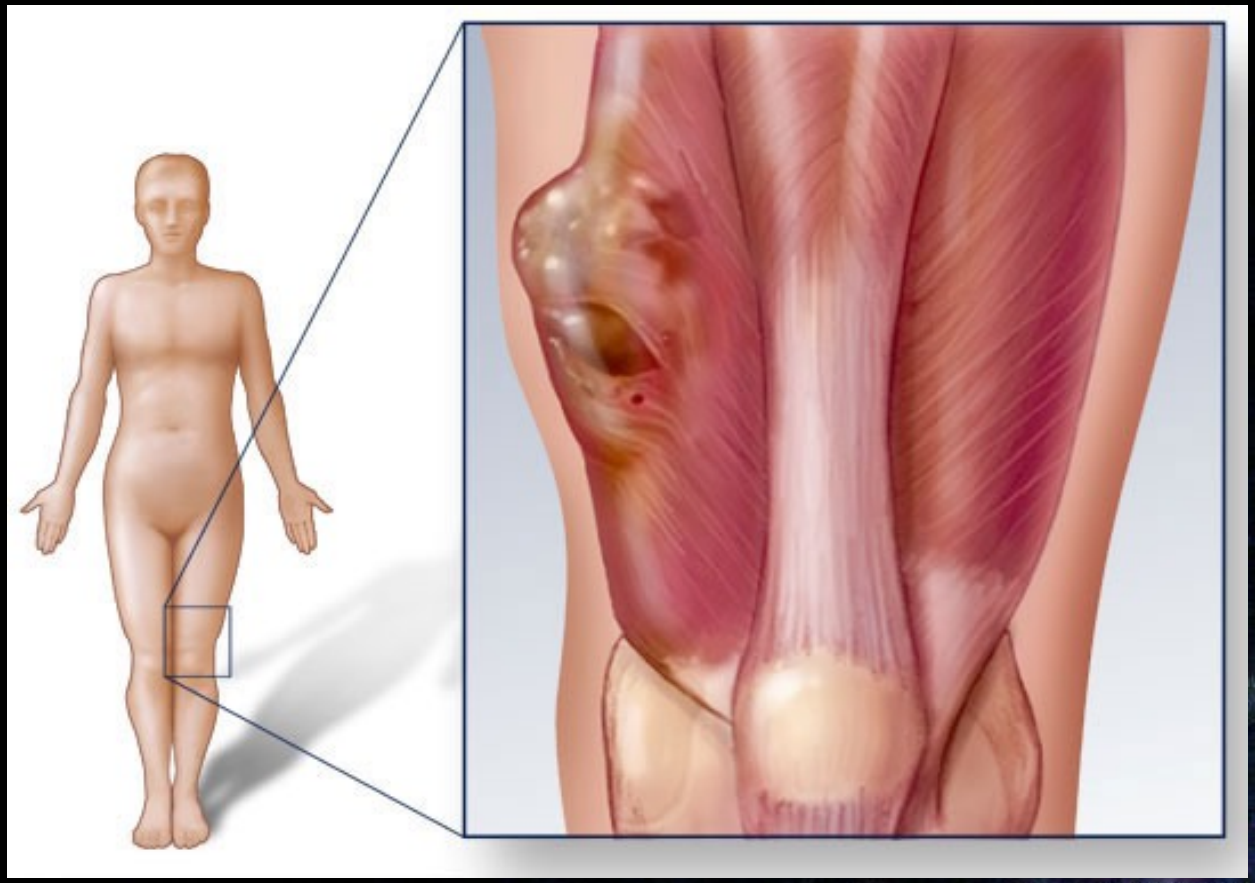
Clinical manifestation

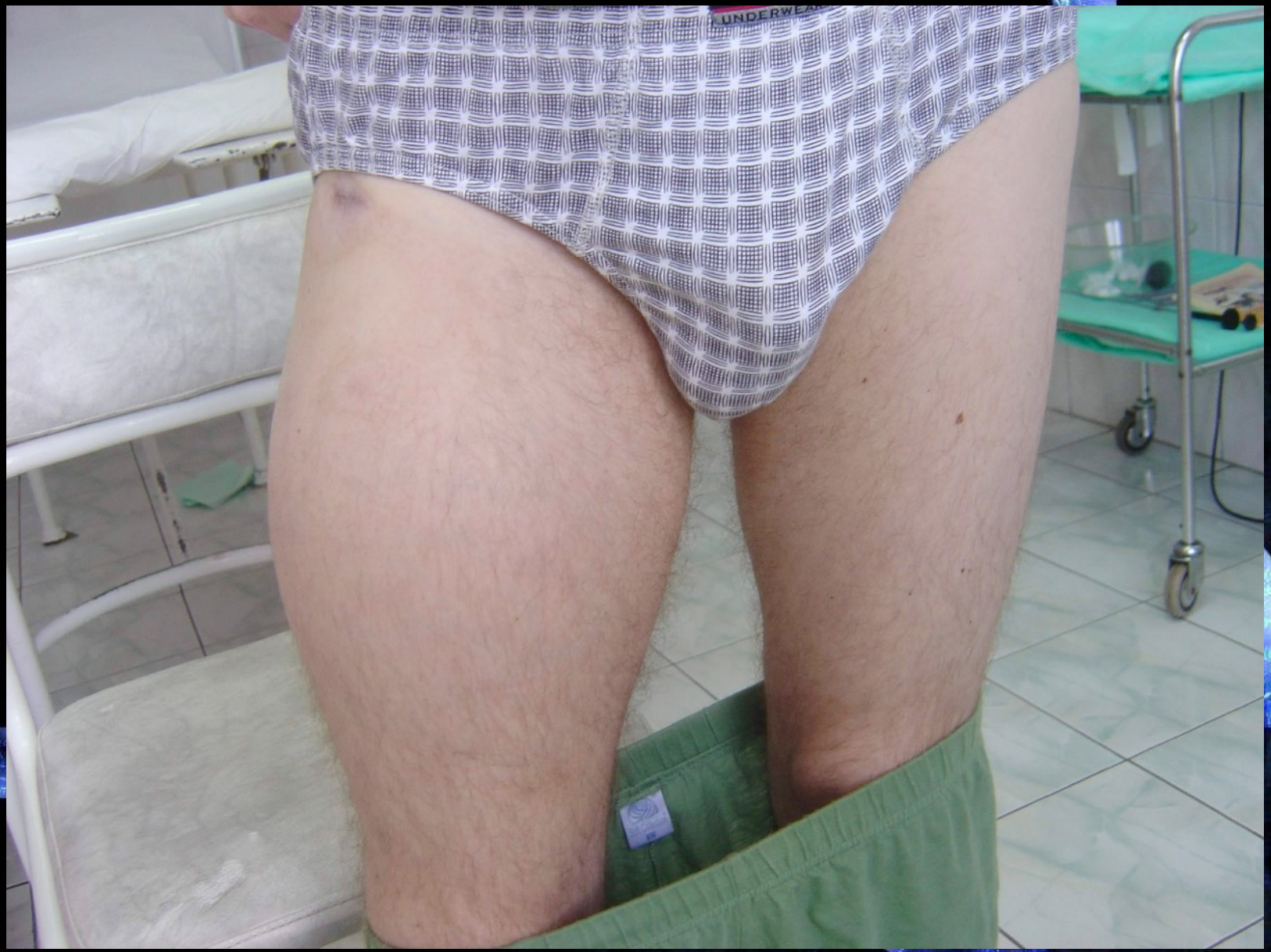
-painless mass
- most common localisation: limbs (especially lower limbs)
-usually bigger than 5 cm
- localisation below the fascia

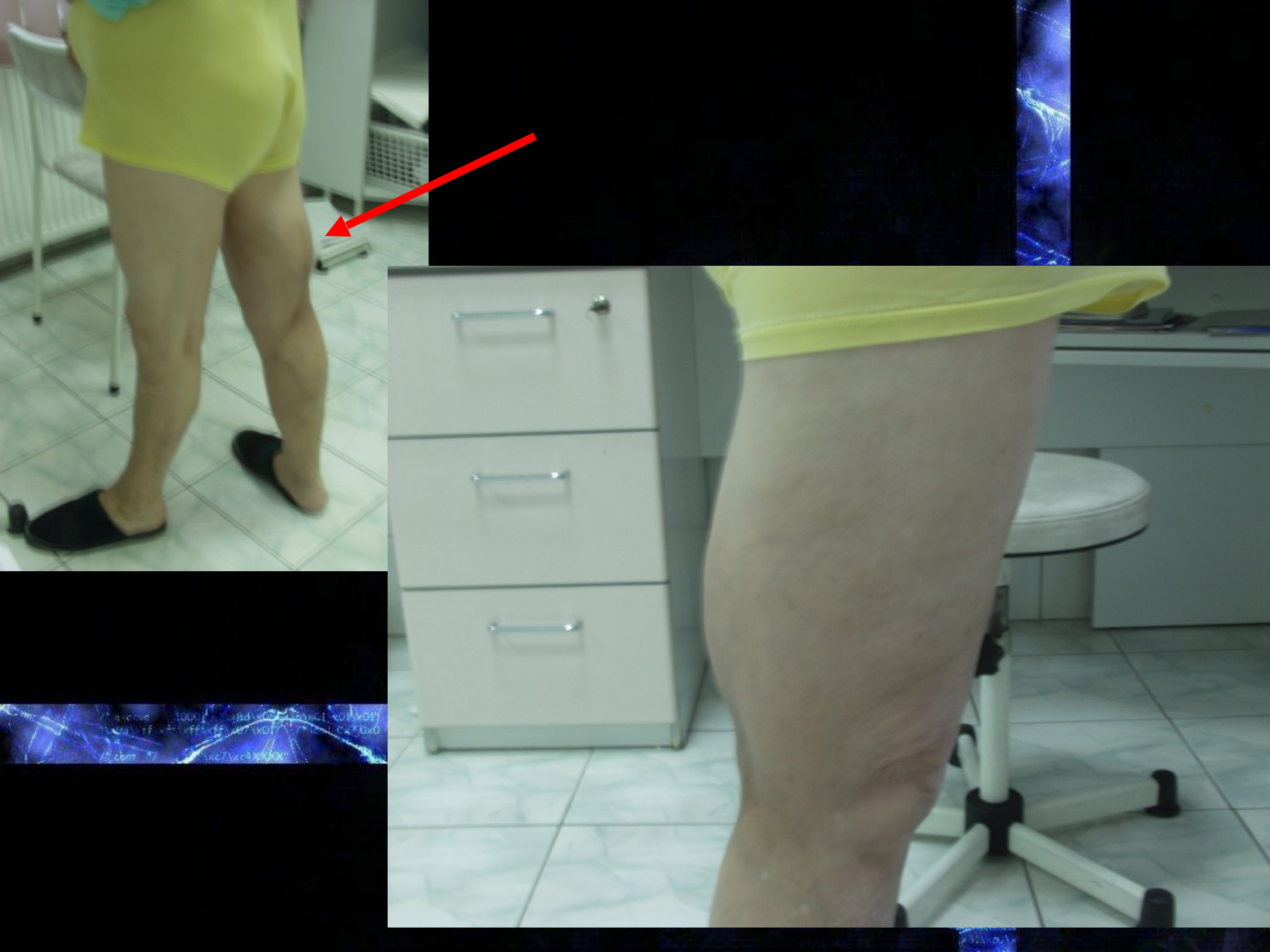
Clinical manifestation

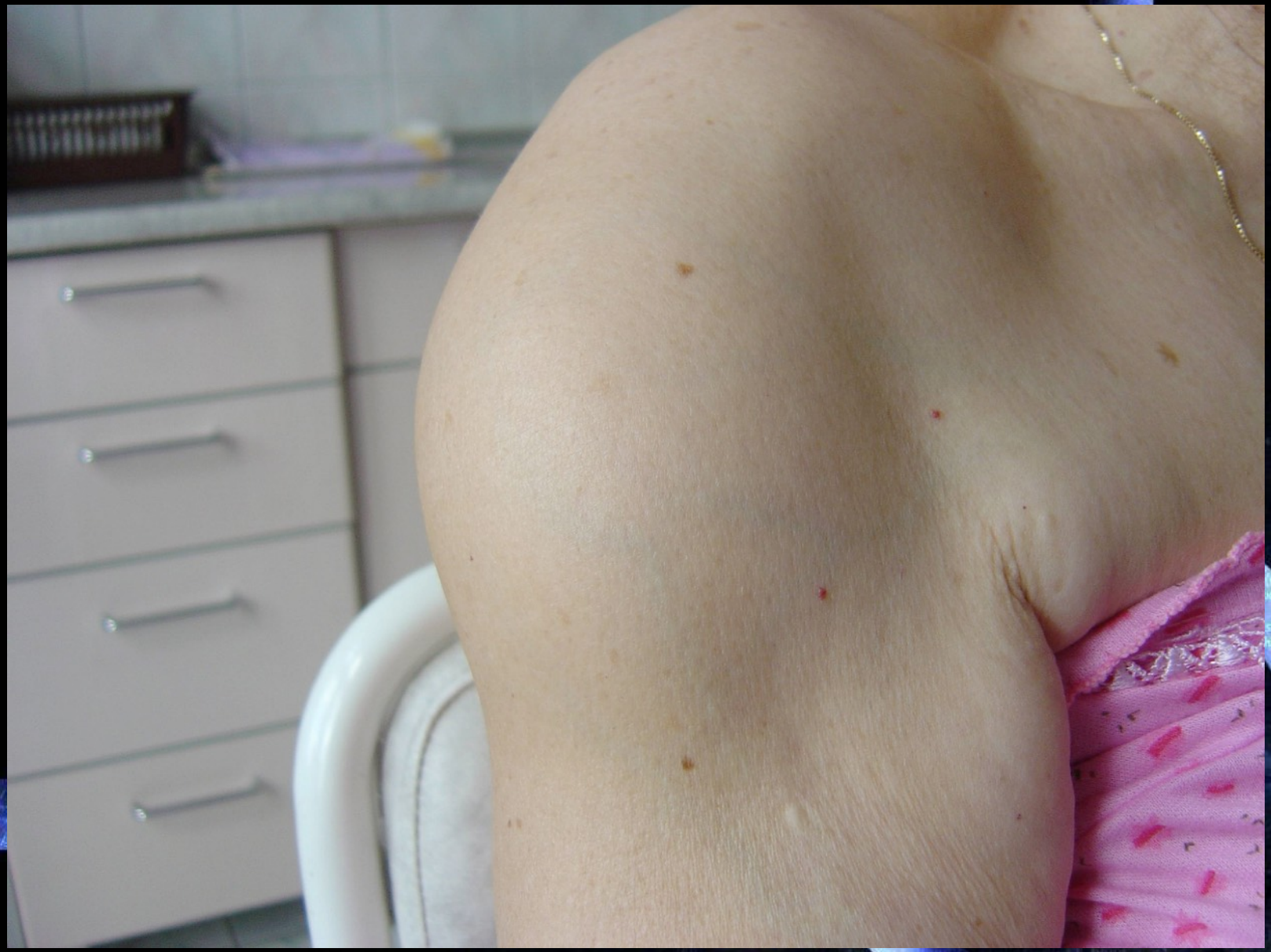
- Rare involvment of neurovascular structures – biologic barriers (although possible) –

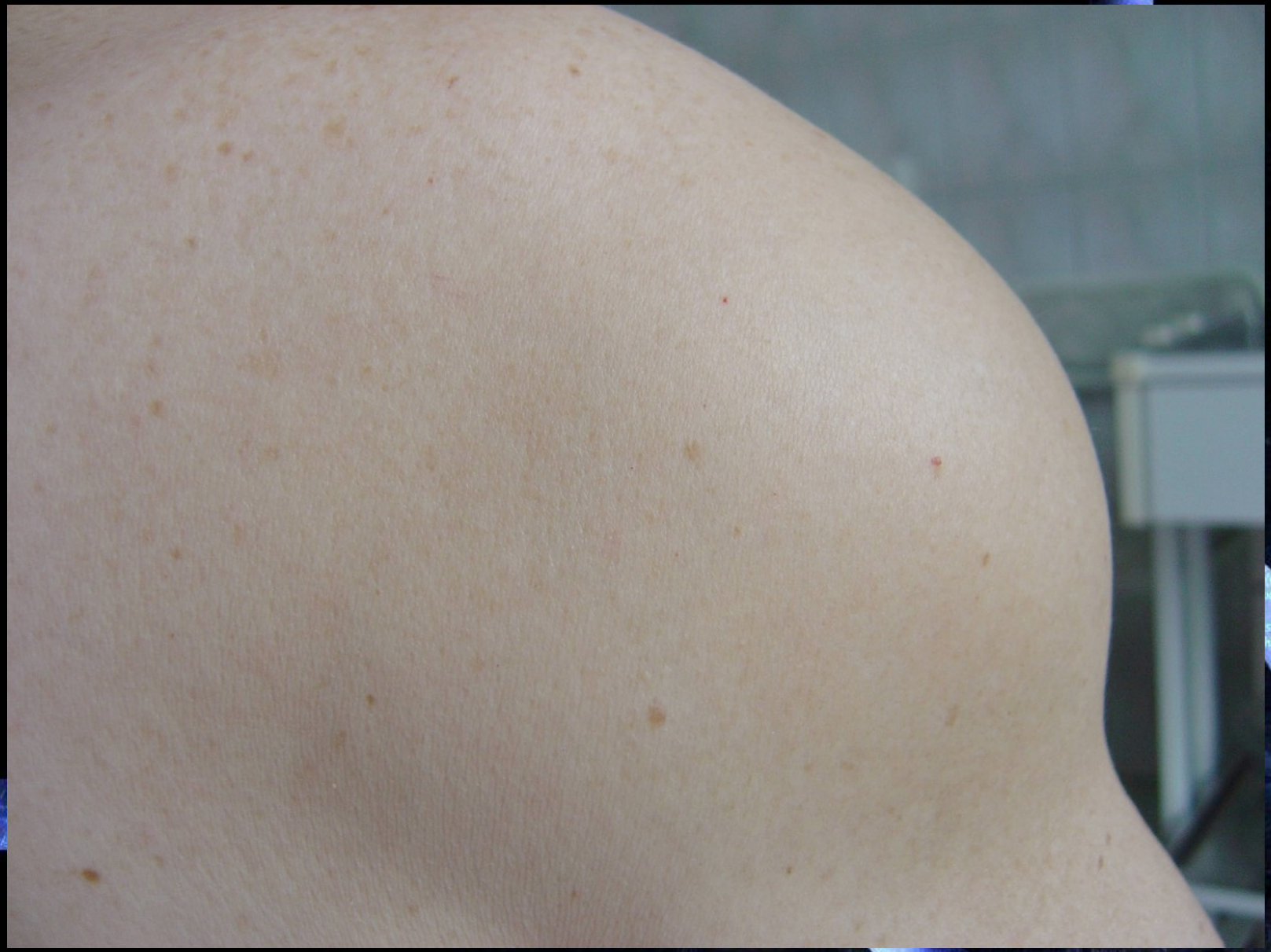
  - usually absence of the neurologic and ischemic manifestation
- Paraneoplastic manifestation – anaemia, hypoglycemia at 2- 4% of the cases

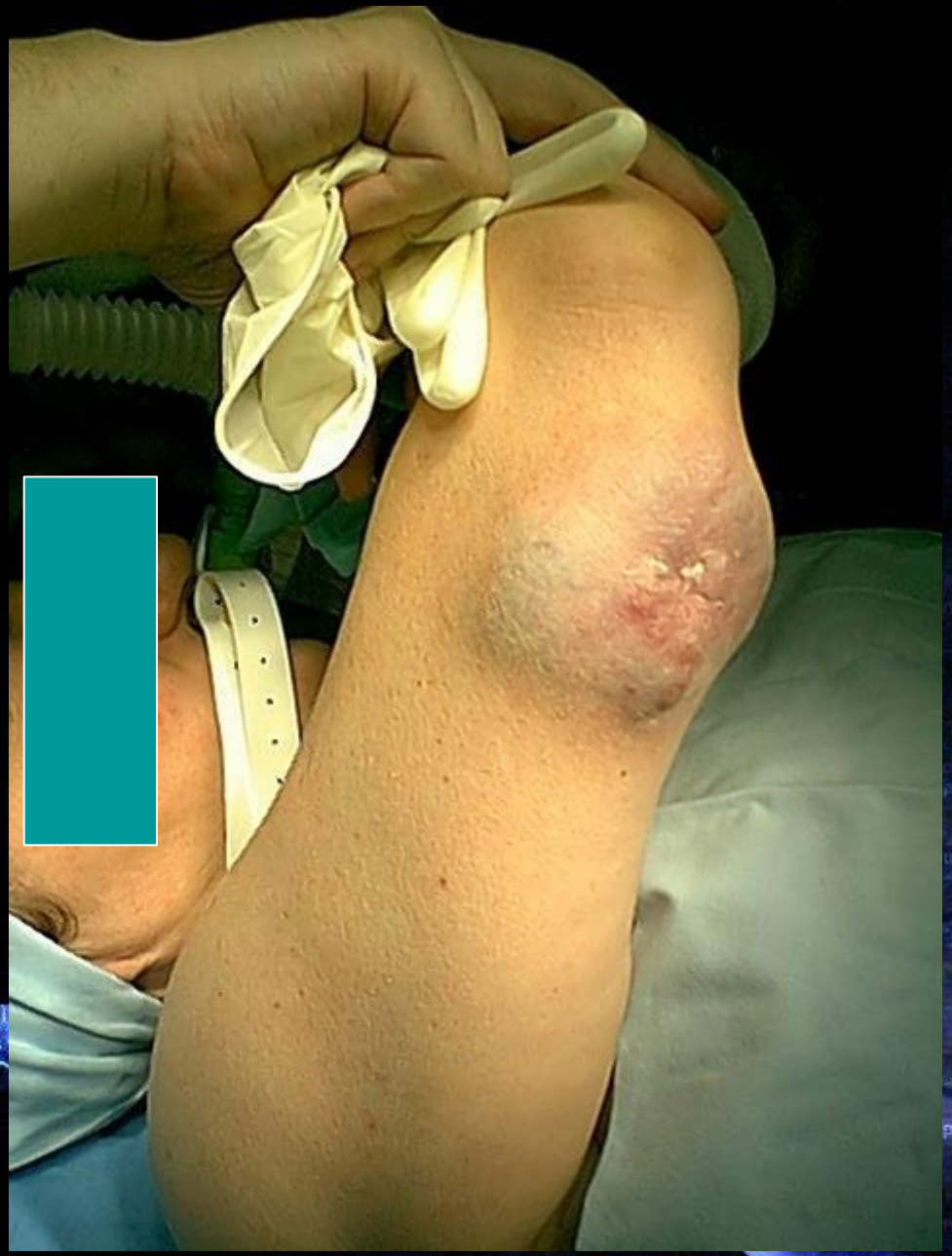


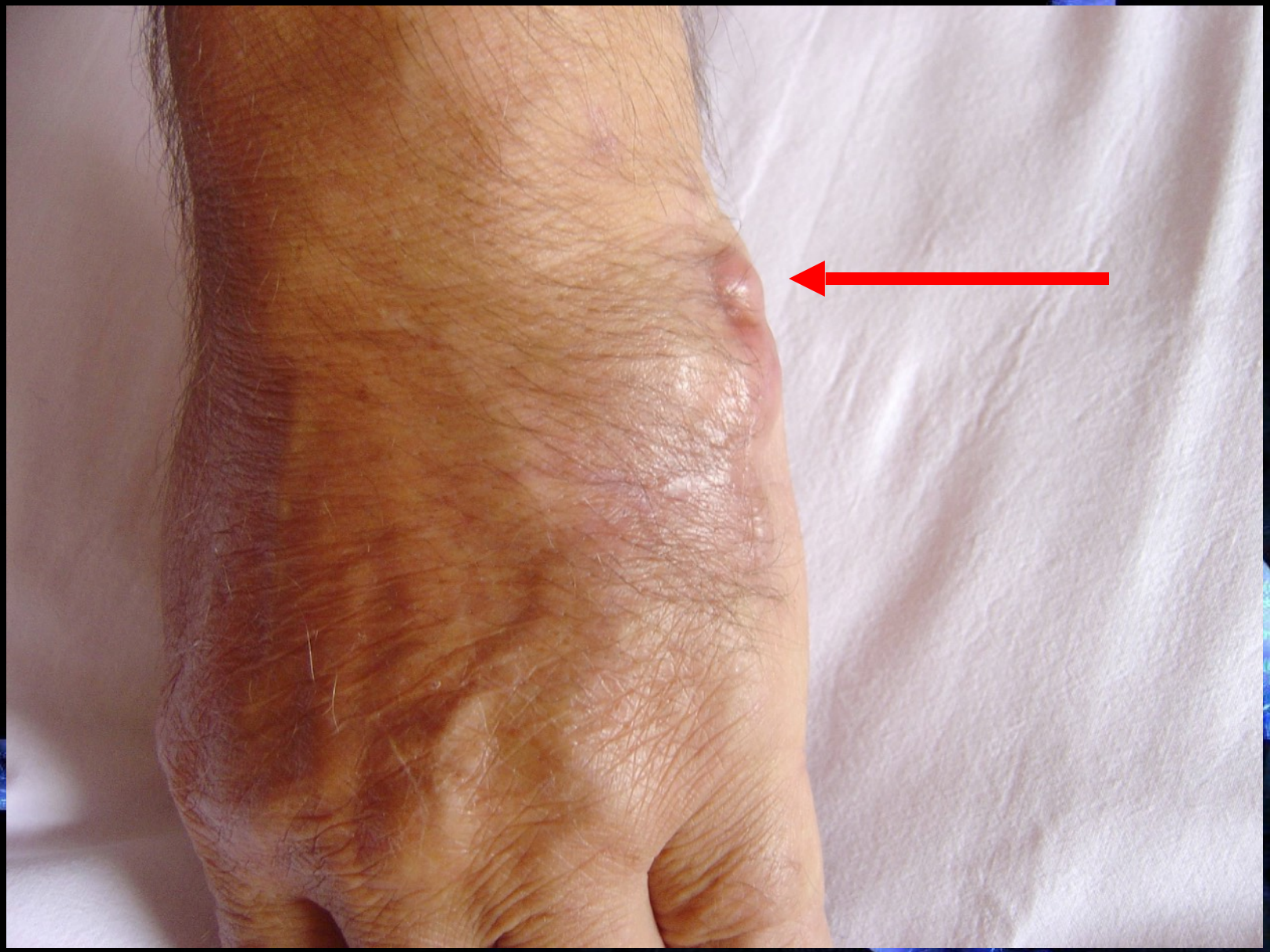


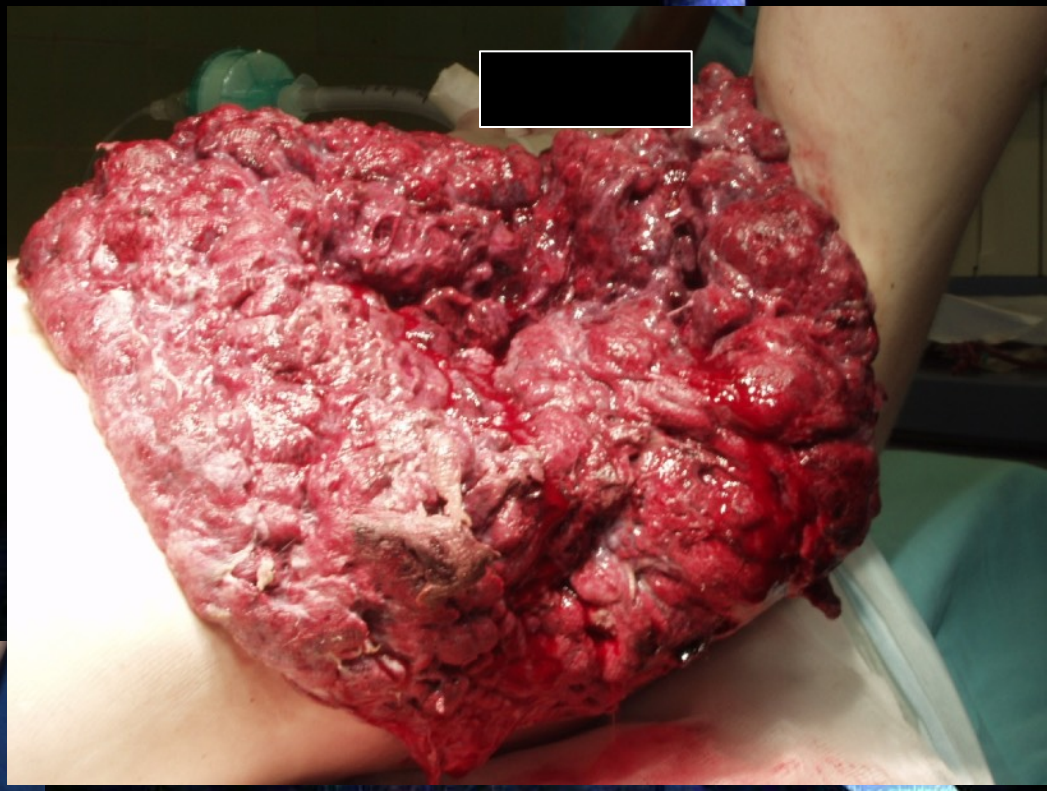


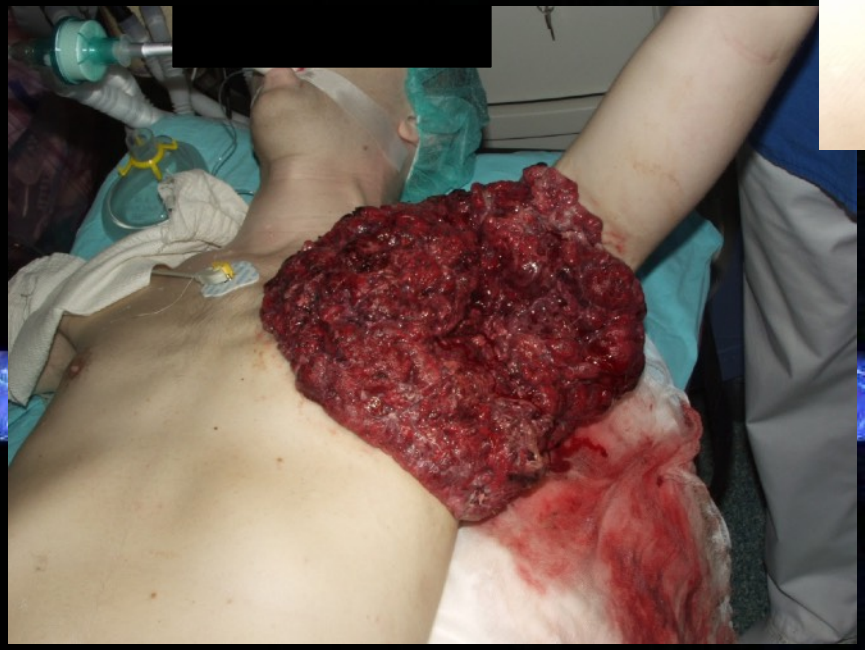


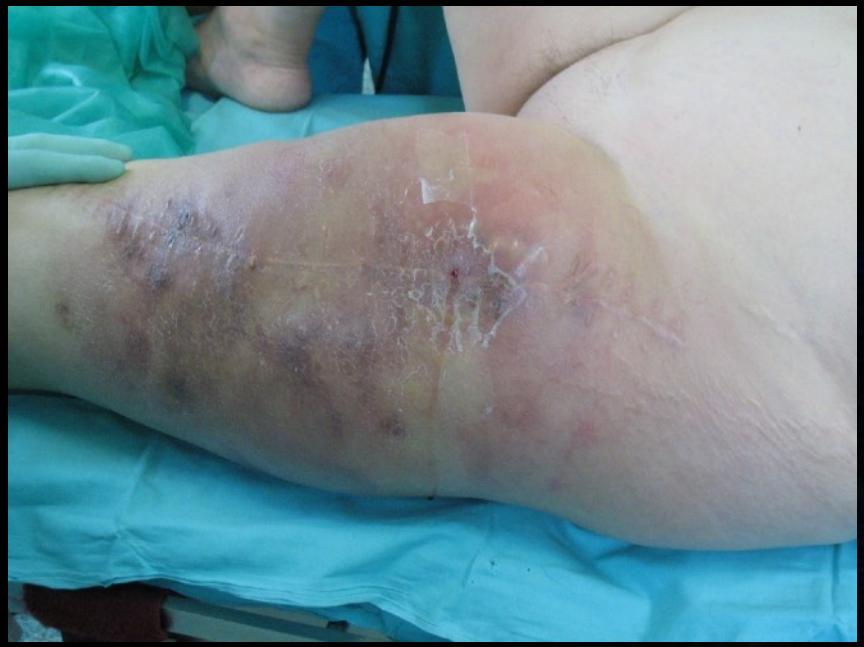


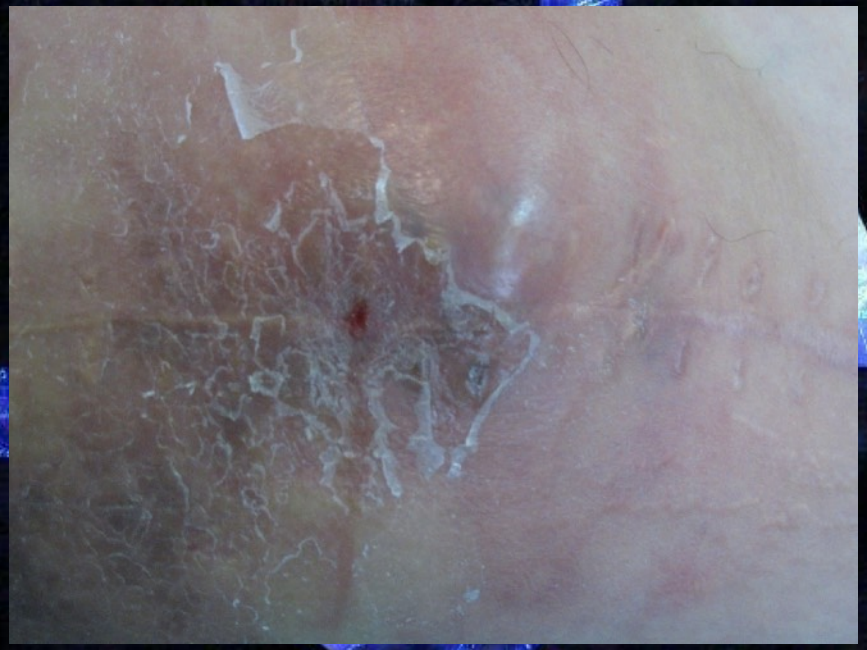


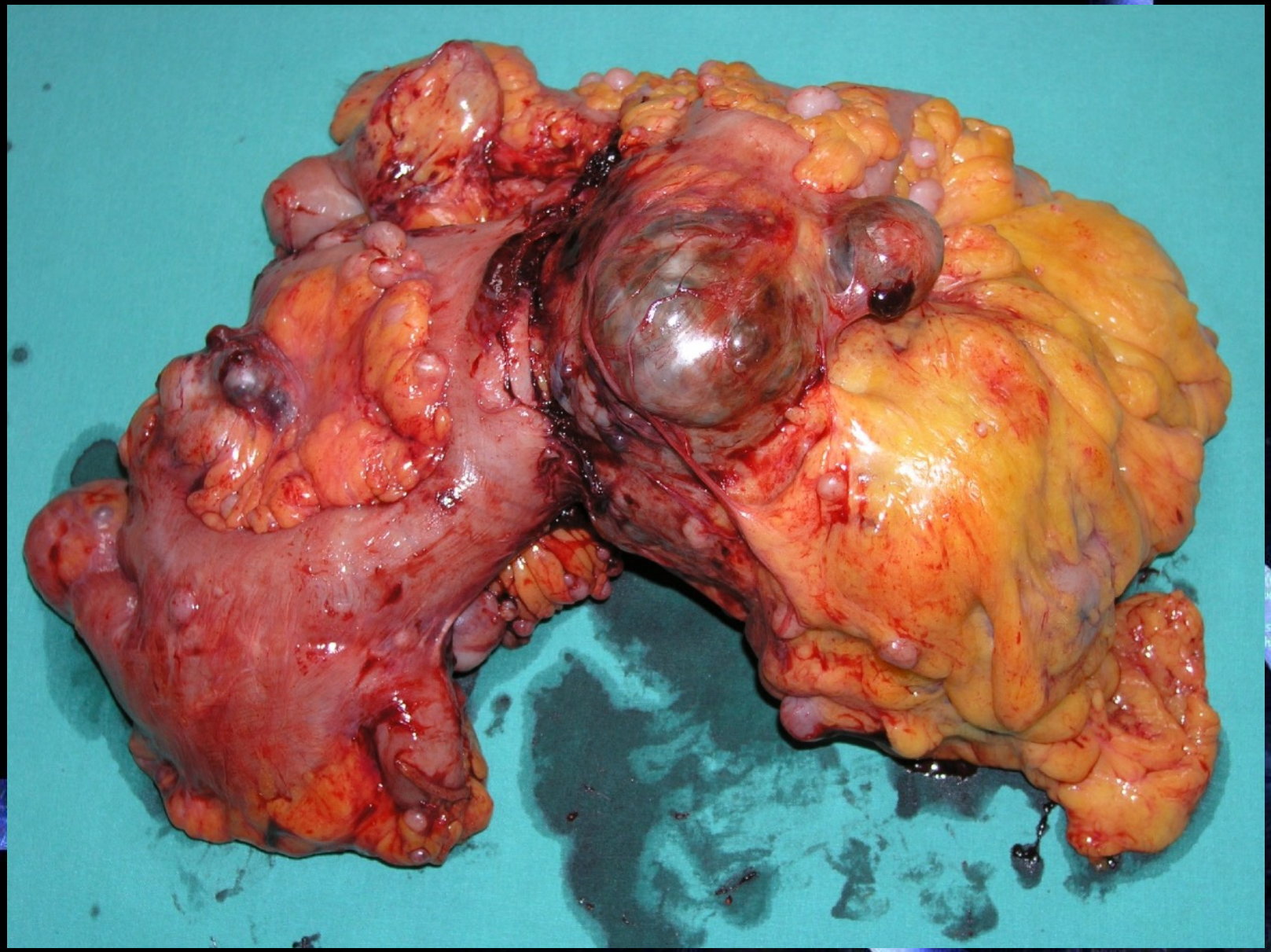












Most common soft tissue sarcomas

1. *Liposarcoma* (tłuszczakomięsak):
>50 yrs, lower limb + retroperitoneal space

2. MFH- *malignant fibrous histiocytoma* (włókniak histiocytarny): 50-70 yrs, tigh, bad prognosis

3. *Rhabdomyosarcoma (2-5 yrs) and leiomyosarcoma (women* >60 yrs, retroperitoneal space, uterus, GI tract)

4. *Sarcoma synoviale* (mięsak maziówki):
big joints, children, bad prognosis

5. MPNST- malignant peripheral nerve sheath tumor- Schwannoma malignum: adults, bad prognosis

Differential diagnosis

-benign soft tissues lesions

(i.e. lipoma, fibroma, angiomyolipoma)

- myositis ossificans

- posstraumatic lesions (hematoma, swelling)

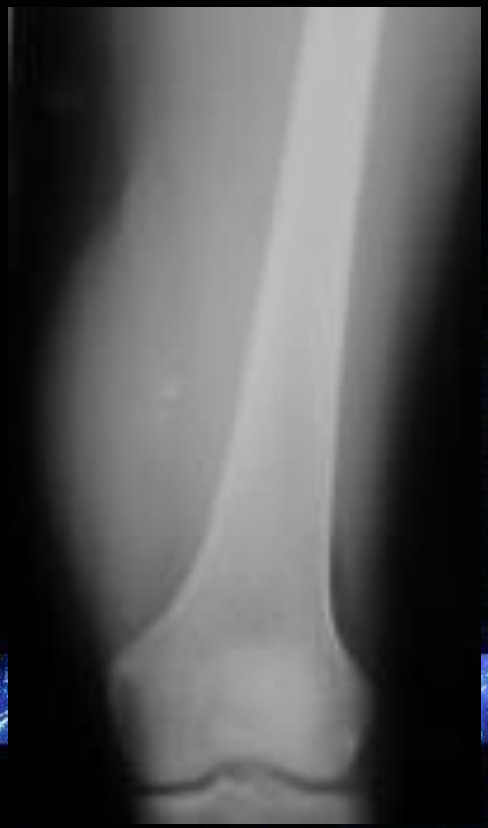
Imaging studies:

- palpation (losalisation under the fascia and mobility)
- X-rays
- ultrasound
- CT
- MRI

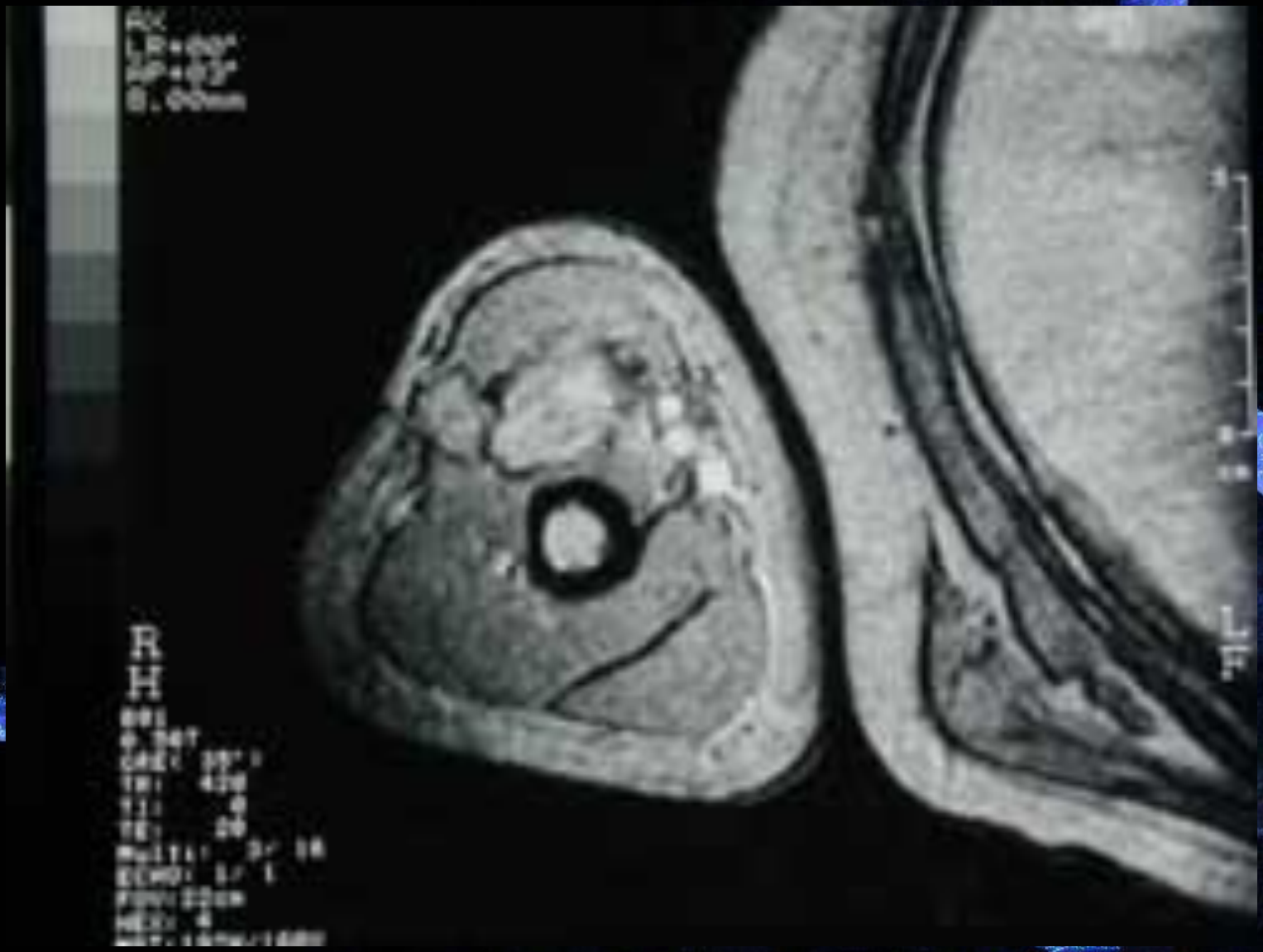
chest X-ray examination, CT of the chest, abdominal ultrasound for intraabdominal lesions

and **BIOPSY**

X-ray



TK



NMR

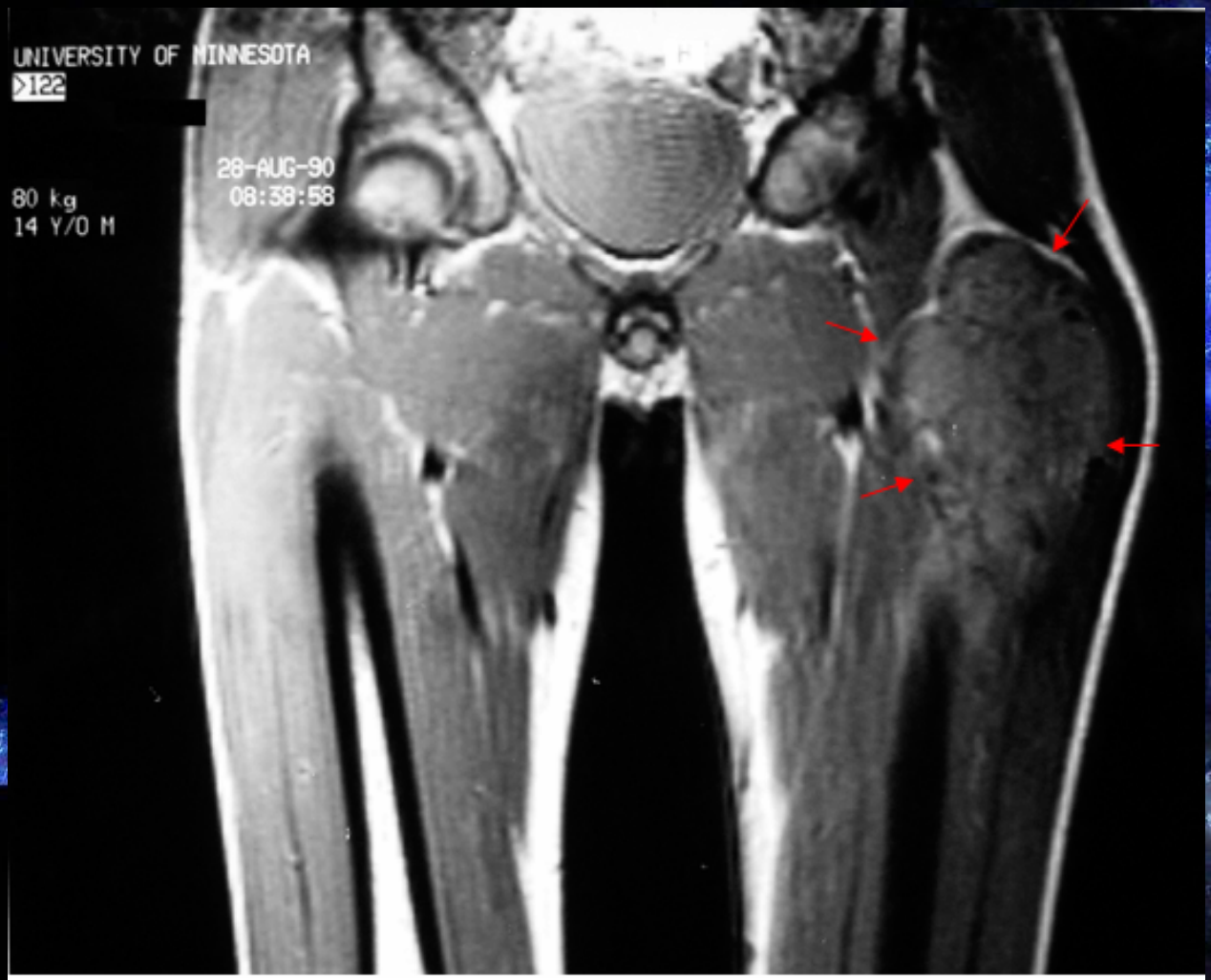


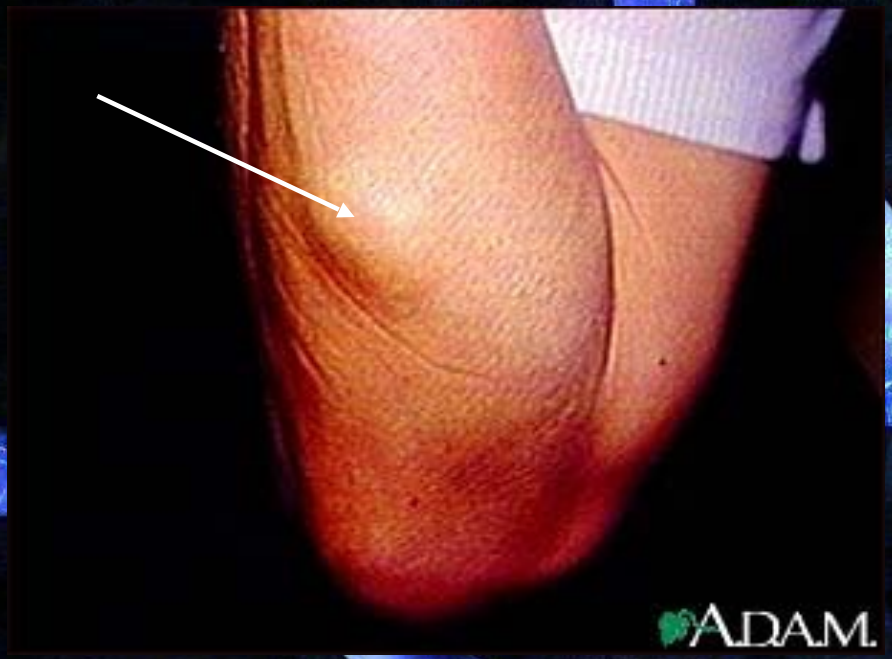
Figure 2 MRI (magnetic resonance imaging) scan of femur with Ewing's sarcoma

Biopsy

- OBLIGATORY IN ALL CASES
- definitive pathologic confirmation as a gate to invasive surgery

Excisional biopsy

- Only in small (less than 5 cm) lesions superficially localised



Biopsy in all other cases (suspicion of sarcoma)

- Fine needle aspiration – not recommended

- Tru – cut biopsy

  or

- Incisional, surgical biopsy

- Frozen section not recommended

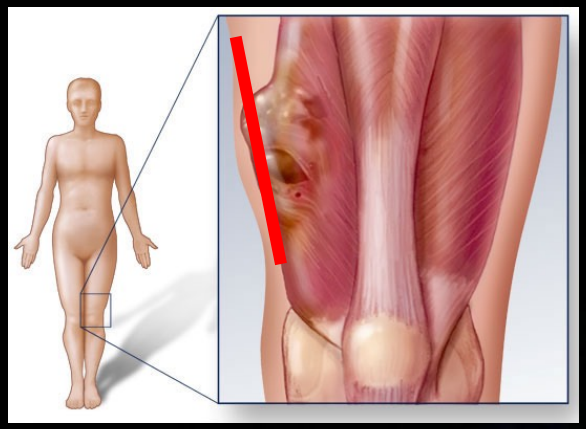
Biopsy

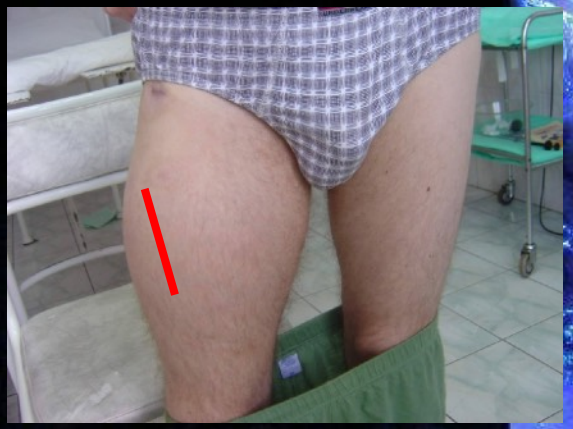
- Obligatory in all cases
- Safe procedure*, does not increase the risk of faillure

  (recurrence and/or dissemination)

* When performed by experienced surgical oncologist armed with imagination

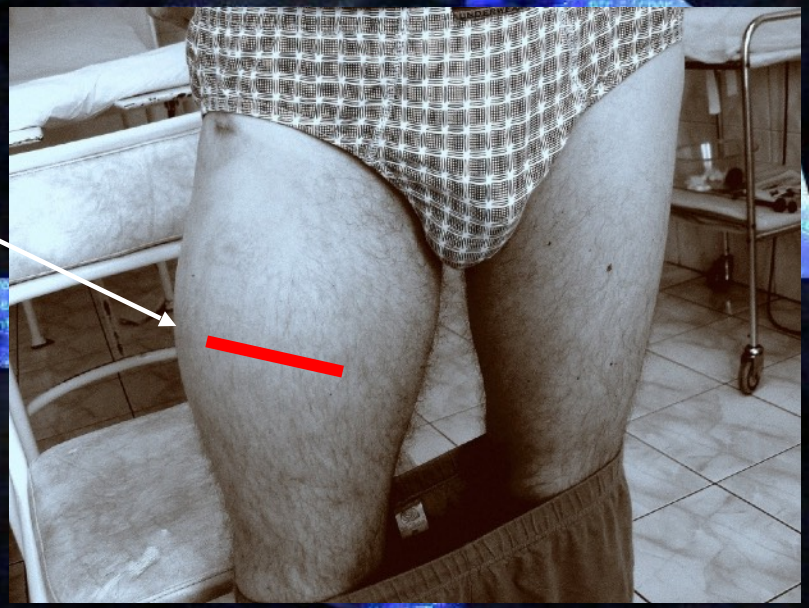
- The biopsy site must obligatory be removed en bloc with tumor during radical surgery





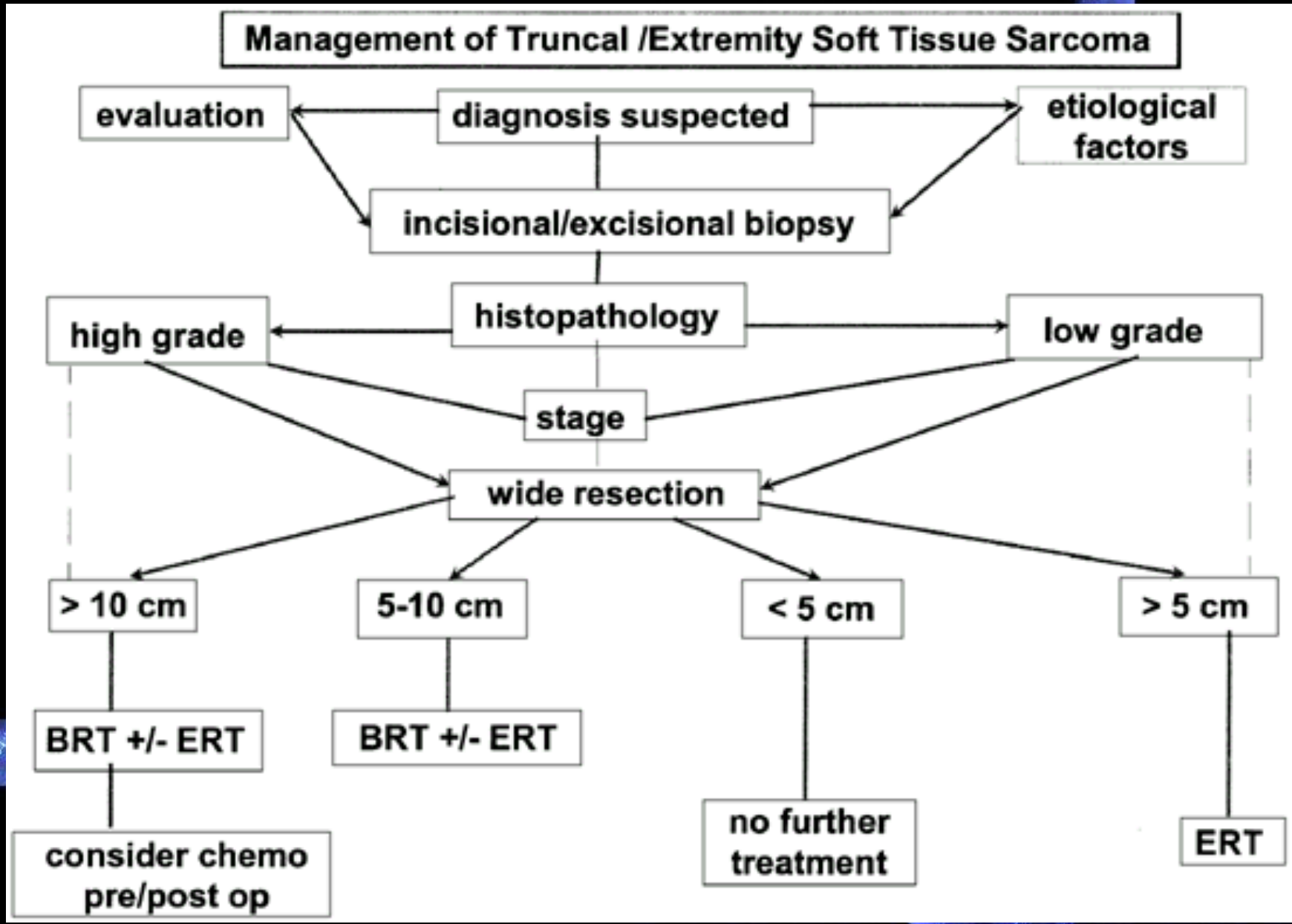
- Right

wrong



The importance of biopsy

- **Mankin et al.. 1982**:
- Serious diagnostic faults 18%
- Lack of material 10%
- AMPUTATION caused by wrong biopsy localisation 4,5%
- Decreased outcome 8,5%


Management of Truncal /Extremity Soft Tissue Sarcoma
evaluation
diagnosis suspected
etiological factors
incisional/excisional biopsy
histopathology
high grade
low grade
stage
wide resection
> 10 cm
5-10 cm
< 5 cm
> 5 cm
BRT +/- ERT
BRT +/- ERT
consider chemo pre/post op
no further treatment
ERT

Treatment: surgery

- radical excision and wide local excision
- caution: pseudocapsule and the tumor's extention beyond it
- the biopsy site and tract taken en block with the tumor/specimen
- regional lymphadenectomy only with cases of the lymph nodes involvment, confirmated with cytologic/ pathologic examination (i.e. FNA)

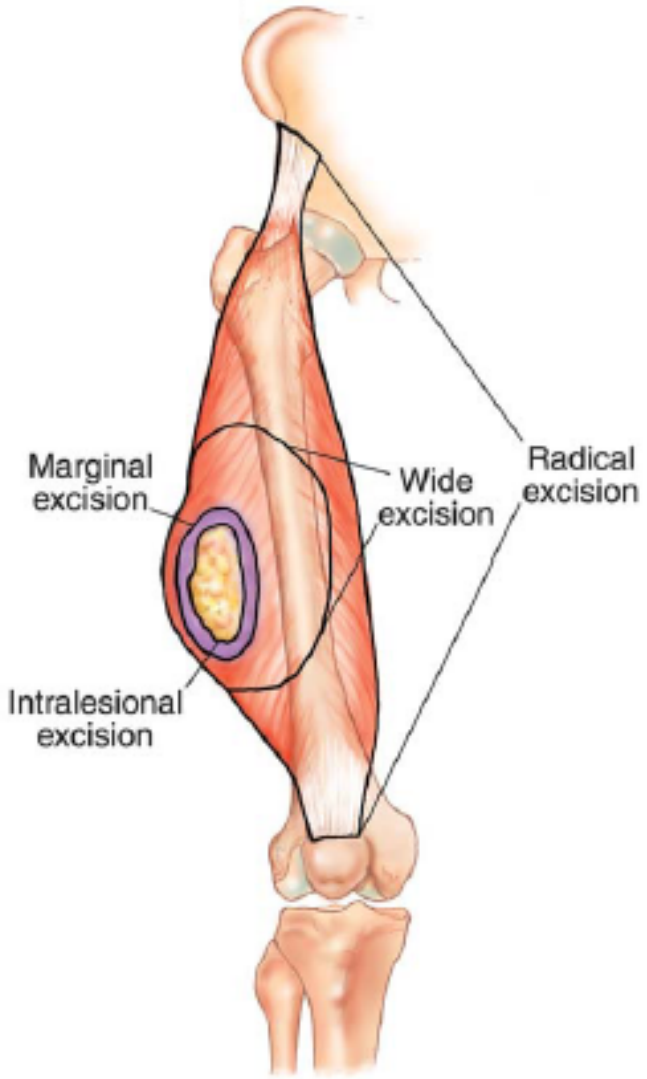


Figure 1.18 Various excision types for soft-tissue sarcoma.

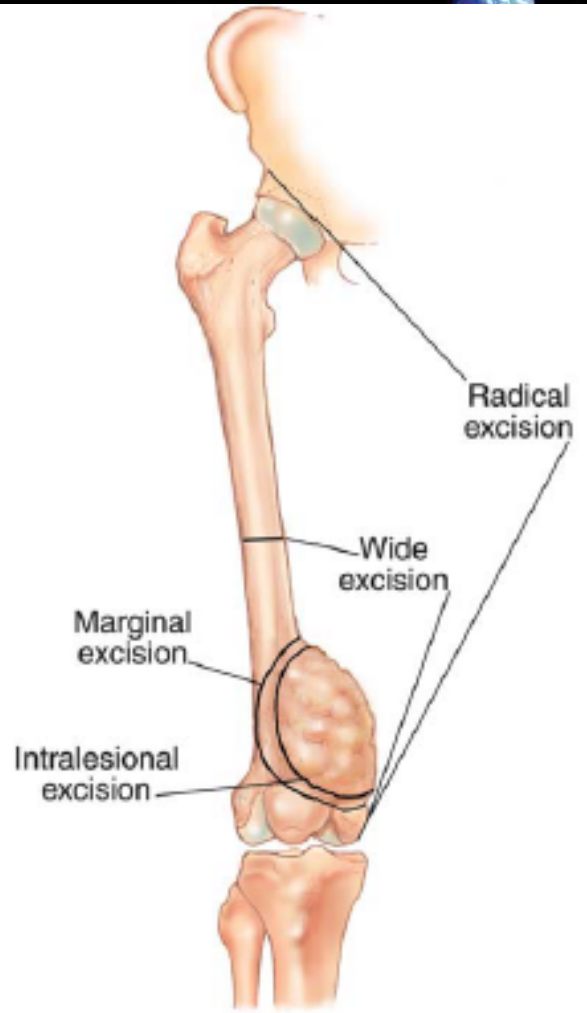
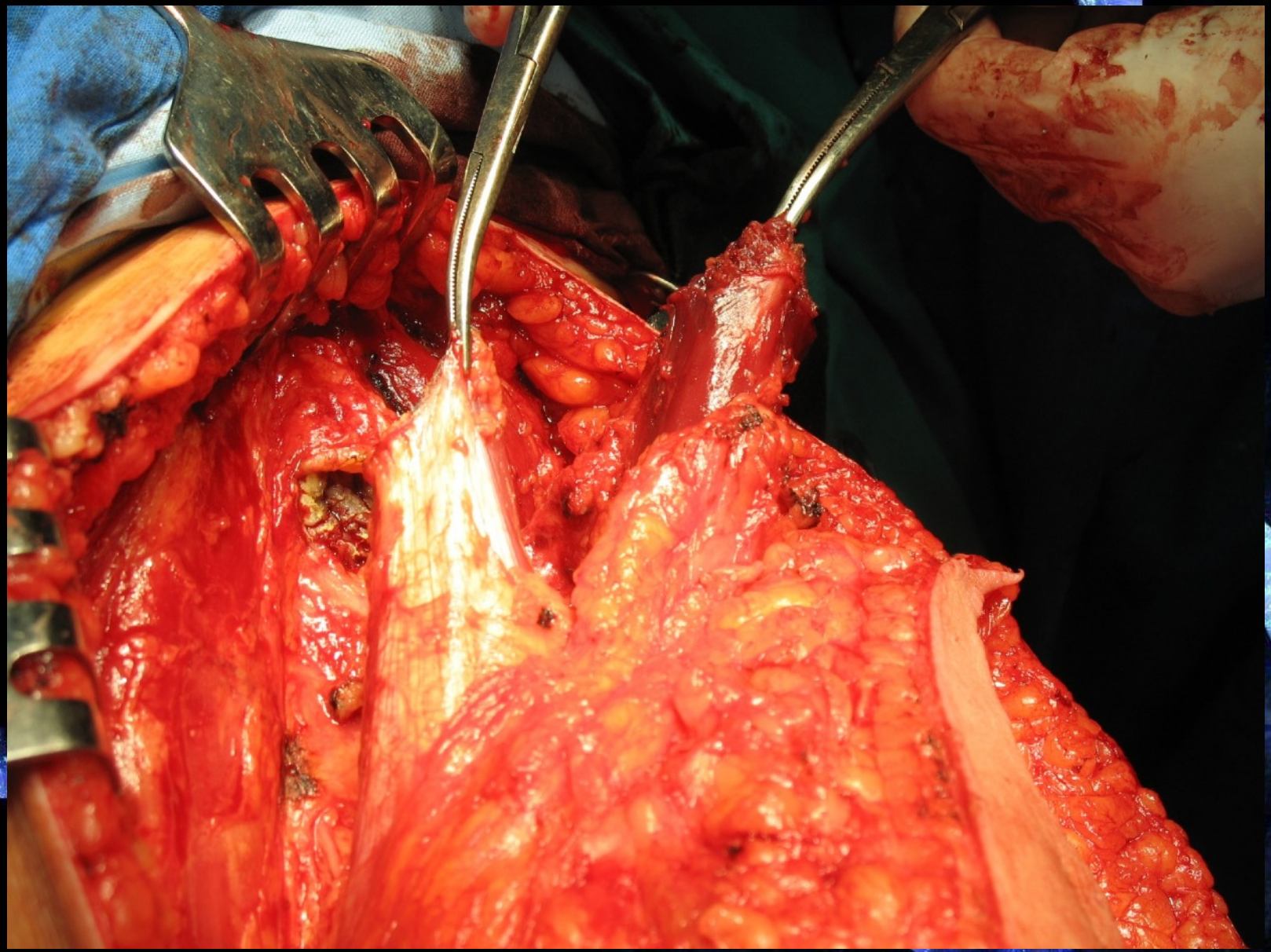
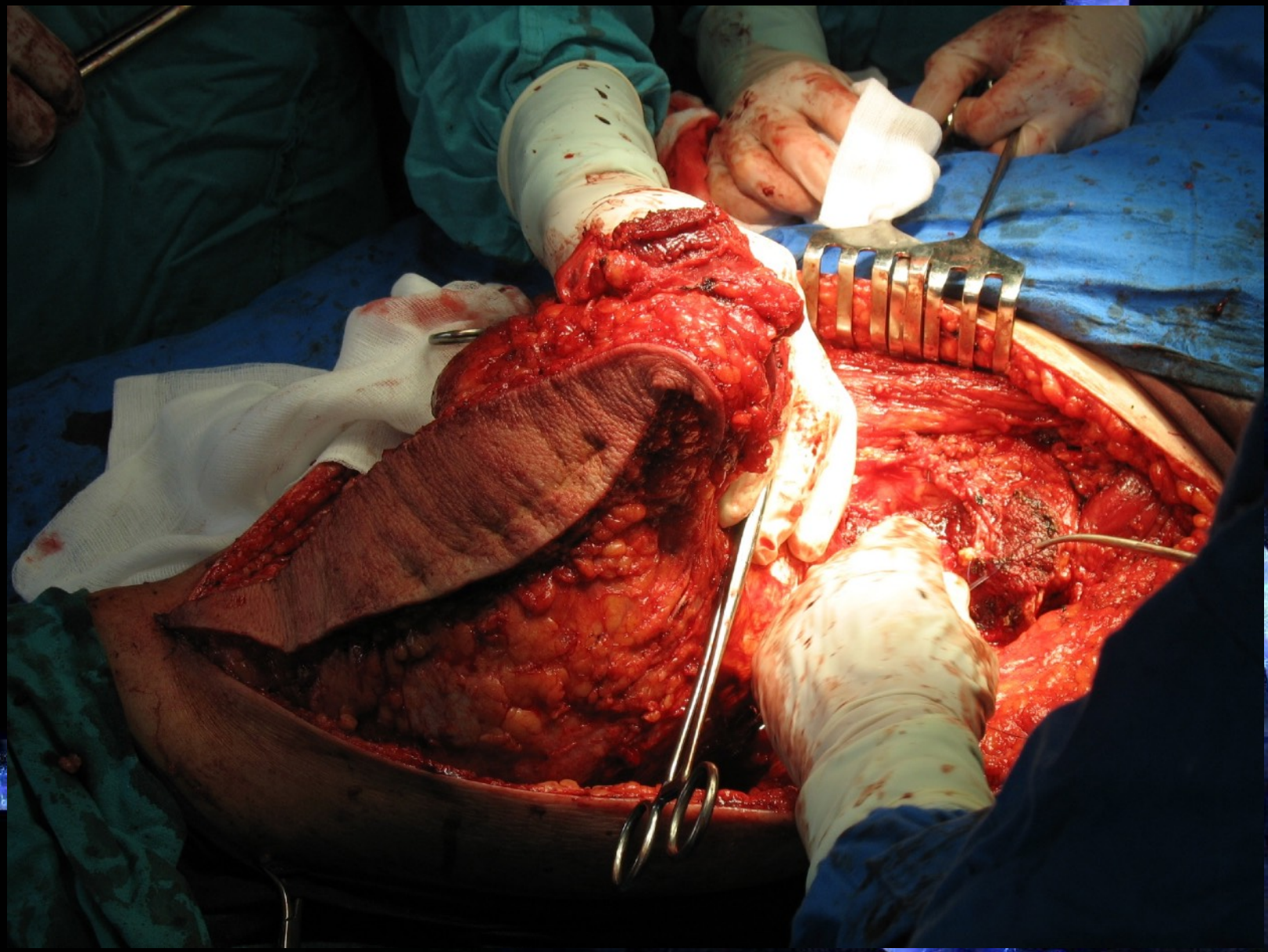
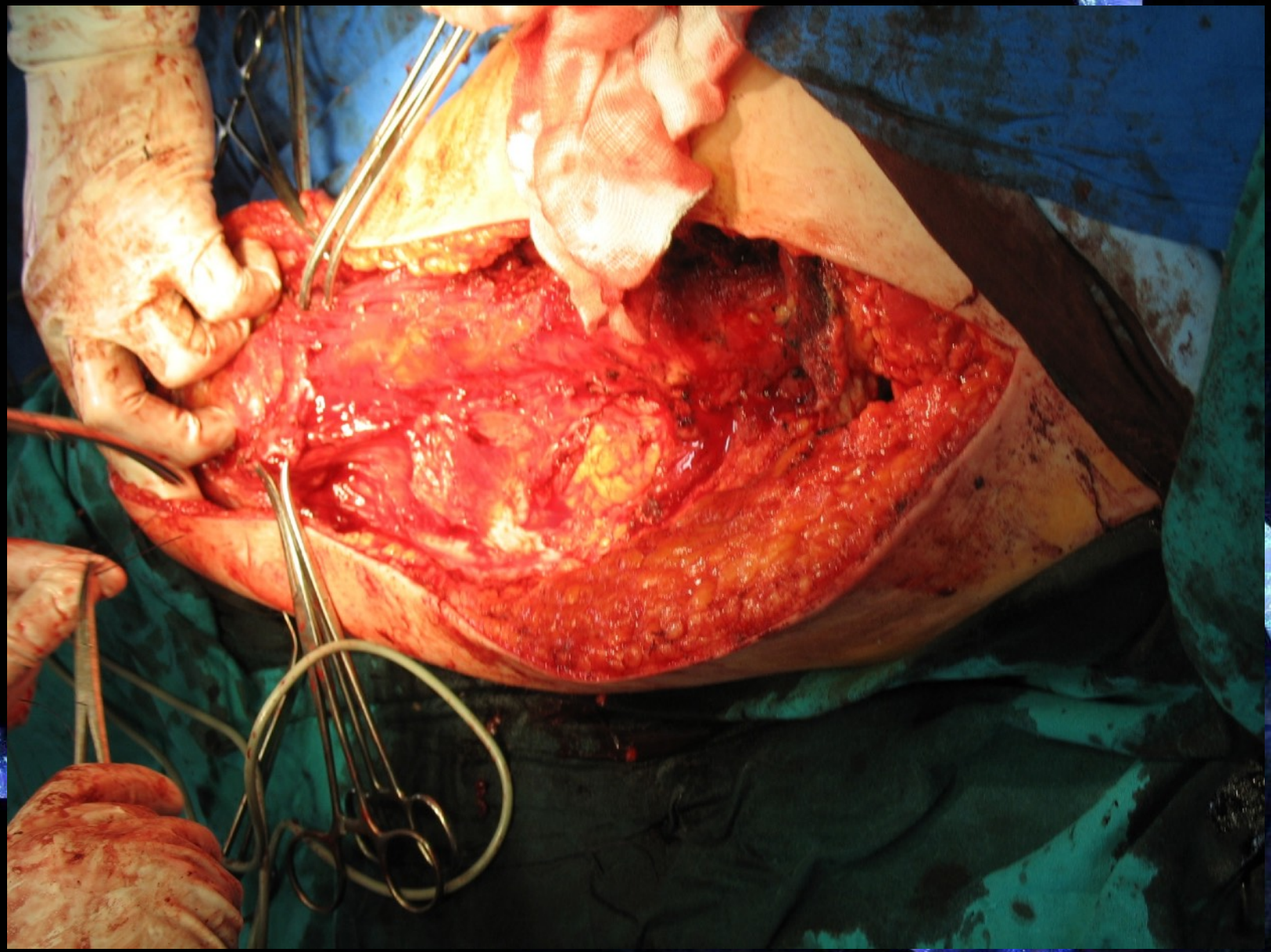


Figure 1.19 Various excision types for bone sarcoma.



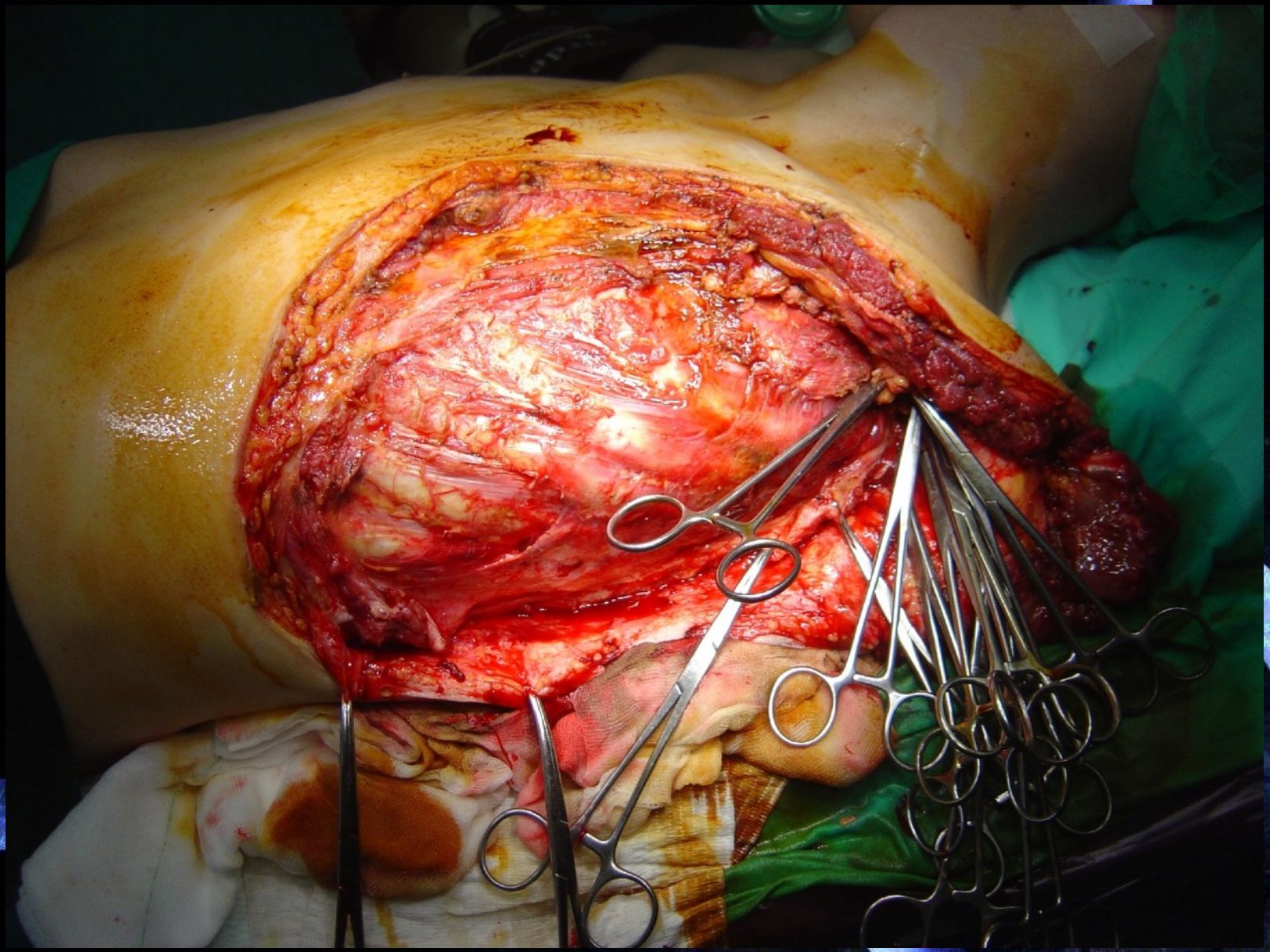




Amputation (indications):

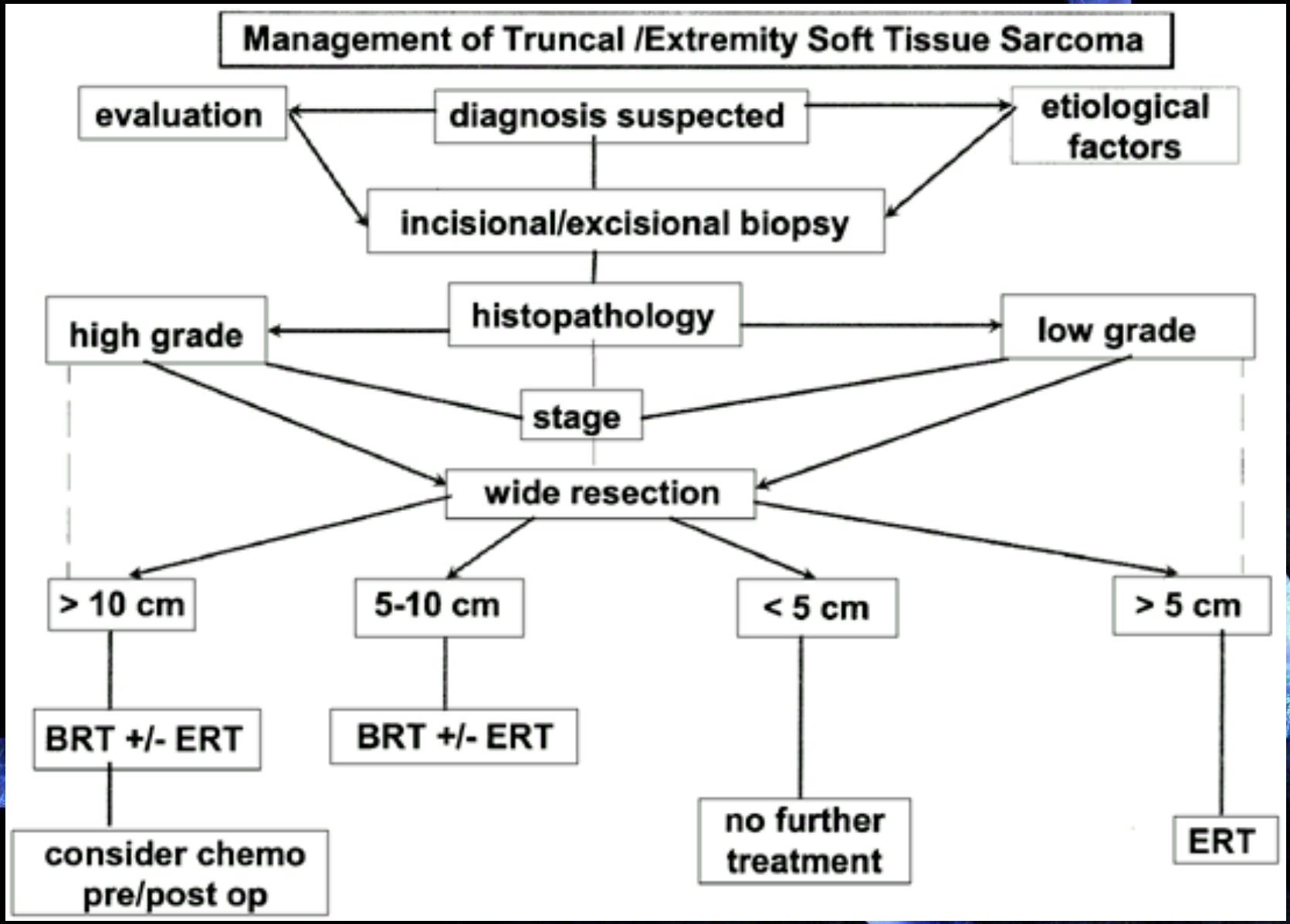
- extracompartmental localisation
- infiltration of the neurovascular trunks
- local recurrence

paliative amputation:
presence of metastases, pain, odour, limb dysfunction



Adiuvant treatment

- Radiotherapy
- Amputation vs limb salvage surgery
- Unsufficient margins
- Head and neck


Management of Truncal /Extremity Soft Tissue Sarcoma
evaluation
diagnosis suspected
etiological factors
incisional/excisional biopsy
histopathology
high grade
low grade
stage
wide resection
> 10 cm
5-10 cm
< 5 cm
> 5 cm
BRT +/- ERT
BRT +/- ERT
consider chemo pre/post op
no further treatment
ERT

Adiuvant treatment

chemotherapy: range of responses 5 – 30%

clinical trials

Most of the sarcomas are rersistant to systemic treatment

place of chemotherapy:

- high-risk localised disease

- metastatic disease

doxorubicine, dacarbazine, isofosfamide

isolated limb perfusion

Trabectedine in liposarcoma

GIST
Gastrointestinal Stromal Tumors

- Stromal tumors of the GI tract
- Uncommon: 1% of the GI tract neoplasms
- Previously classified as leyomyoma and leyomyosarcoma
- Loc.: stomach – 65 – 70%
  small intestine: 25 – 45%

Less frequently: oesophagus, colon and rectum

GIST - treatment

- Combined modality treatment:
  Surgery (R0 margin) and imatinib
- Lymph node involvment is uncommon

Bone sarcomas

epidemiology

- Malignant tumors of the skeletal system – 0,2% of all cancers (US)
- Osteosarcoma and Ewing's sarcoma are the two most common bone sarcomas.
- Osteosarcoma – frequency peak during adolescent growth
- Ewing's sa – second decade of life

Etiology

predisposing and associated factors:

- Diseases with increased bone metabolism
- Benign bones lesions (exostoses for chondrosarcoma)

Clinical presentation

- Pain and swelling of the bone and/or joint
- Caution: traumatic event in anamnesis can delay diagnosis
- Pain more intensive at palpation

Clinical presentation localisation

- Osteosarcoma – metaphysis of long bones, especially distal femur, proximal tibia, humerus
- Ewing's sarcoma – flat bones and diaphysis of femur, tibia and fibula.

  Also in soft tissues
- Chondrosarcoma – pelvis, proximal femur

Dissemination pattern

- 25% of the patients have metastases at presentation
- Localisation: lungs – 90%, bones – 10%
- Ewing's sa – 25% metastases in bone marrow

Imaging studies

- X-Ray, plain radiographs
- MRI
- Bone scanning
- X-Ray/CT of the chest

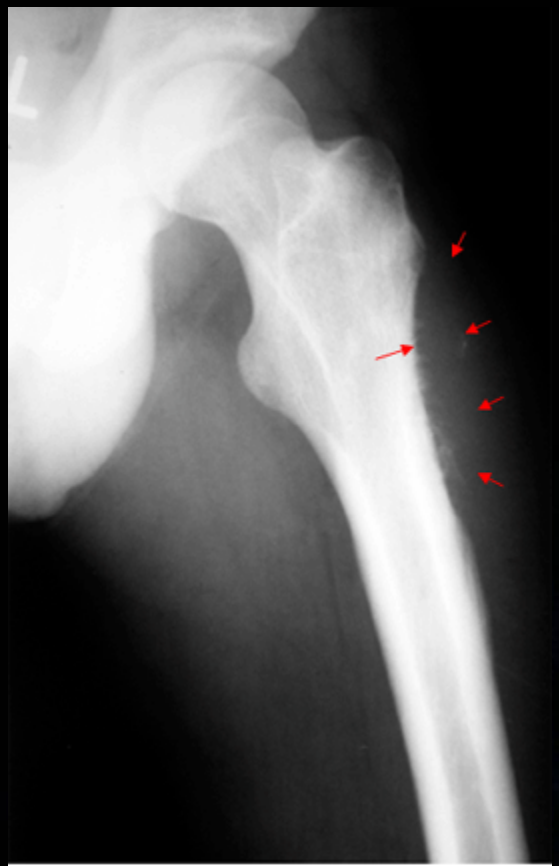


Figure 1 X-ray (radiograph) of thighbone (femur) with Ewing's sarcoma.

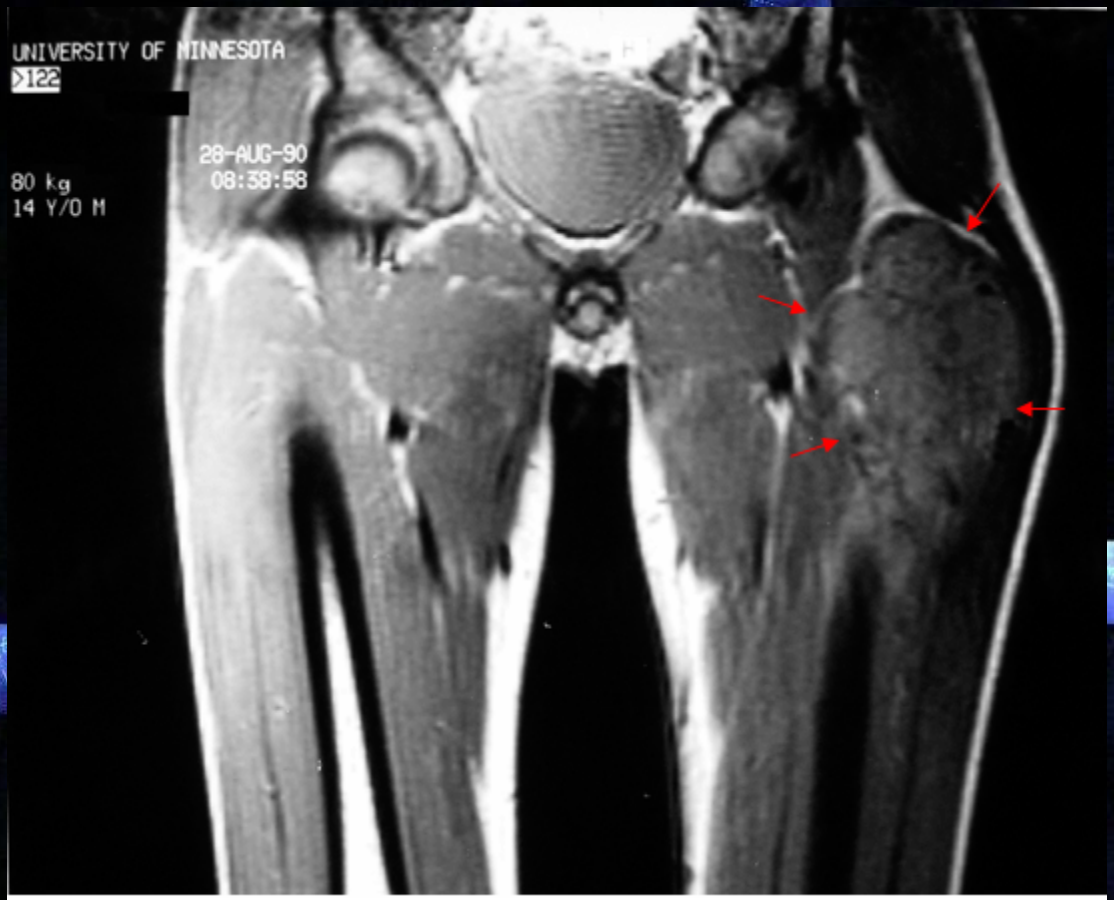
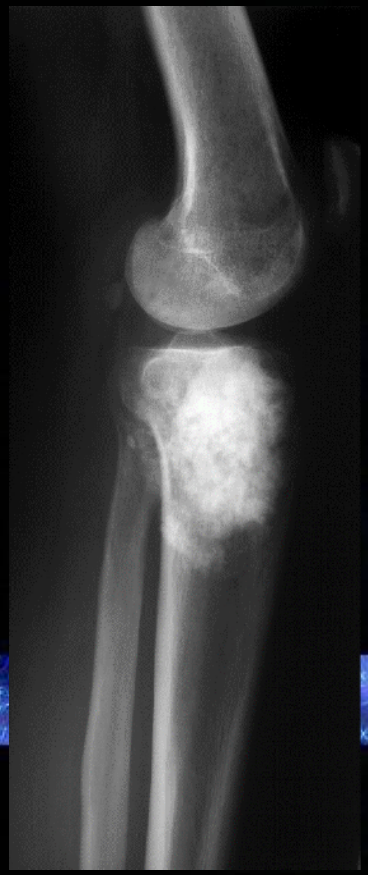
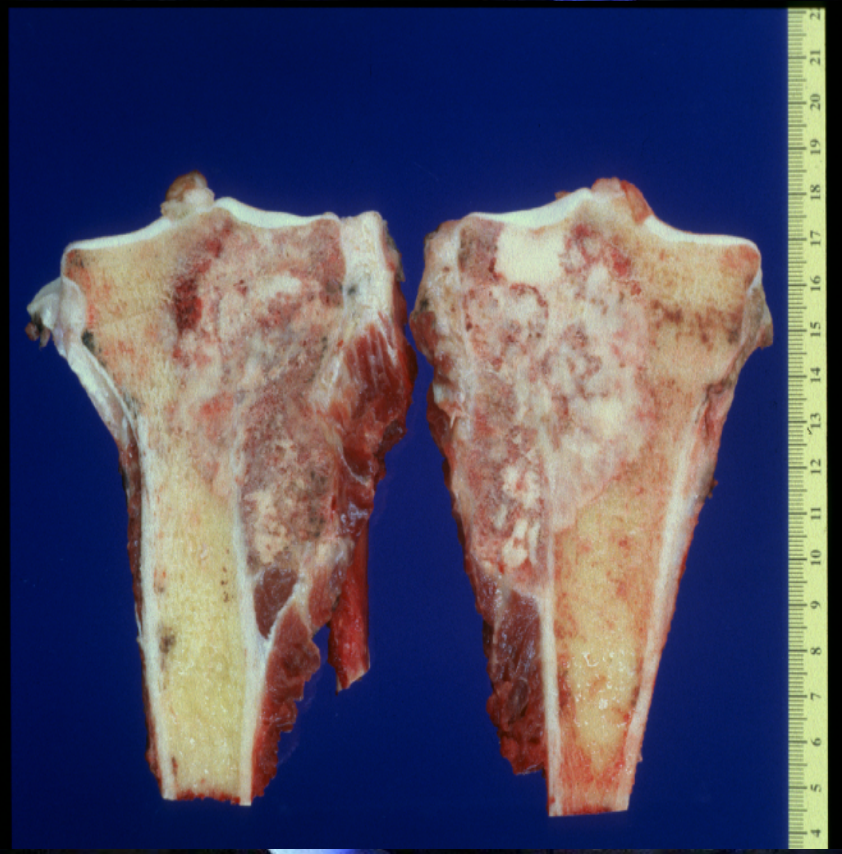
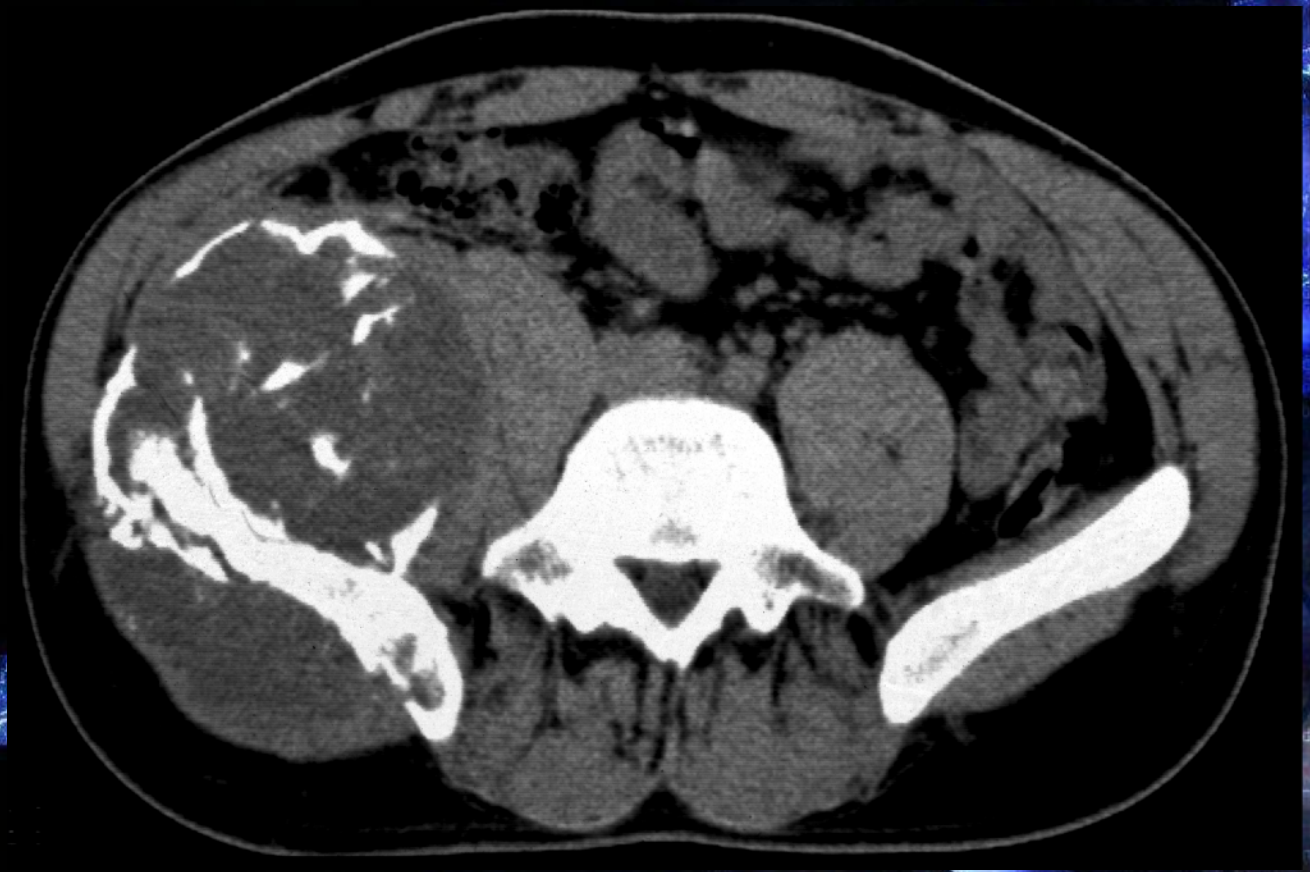


Figure 2 MRI (magnetic resonance imaging) scan of femur with Ewing's sarcoma

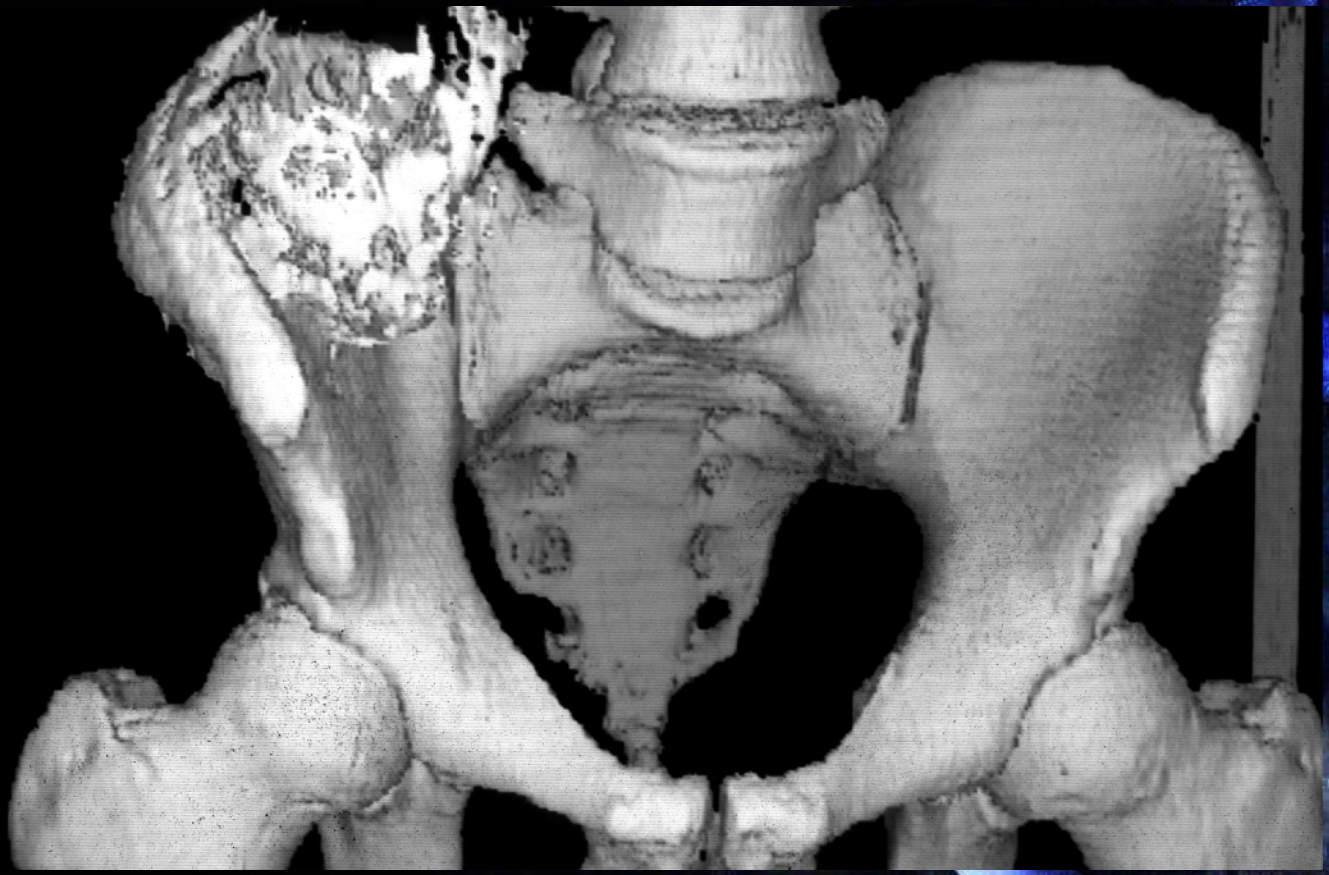




Chondrosarcoma CT scan



3D CT



Biopsy

- OBLIGATORY IN ALL CASES
- definitive pathologic confirmation as a gate to invasive surgery
- Core needle biopsy and incisional/surgical biopsy

osteosarcoma

- Biopsy, evaluation
- Chemotherapy as a standard of care

  15% OS - surgery alone vs 75% OS with combined treatment
- Limb sparring surgery: tumor resection, bone reconstruction, soft tissue coverage
- Limb function
- Postoperative chemotherapy
- Resistant to radiotherapy
- Pulmonary metastasetomy

Bone sarcomas

- **spindle cell**

Oseosarcoma, chondrosarcoma, tumor gigantocellulare

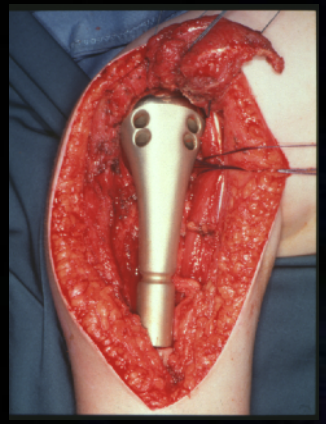
- **small cell**

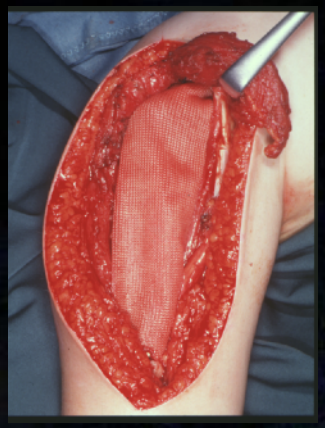
Tumor Ewinga, angiosarcoma, myeloma, PNET

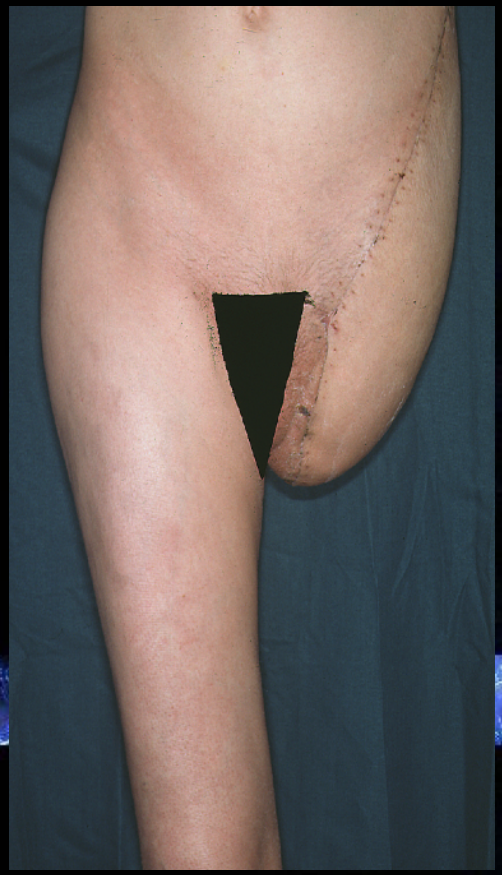
Ewing's tumor

- Age: 5 a 25 r.ż.
- 60% males
- Localisation: limbs – 50% pelvis - 25%, vertebral column and scapula - 15 %
- Pain
- Chemotherapy and radiotherapy, but:
- Surgical excision of the tumor is favorable

Thanks to dr-a Tunna
Robert Roessle Klinik, Charite, Berlin







PROGNOSIS

♦the outcome depends on the type of sarcoma, histopathological grading, the response to adjuvant CHTH

♦ bad prognosticators are: grade G3 and 4, large tumor dimension, localization different than the extremities, meta in bone marrow, treatment of the primal focus at the stage of progression