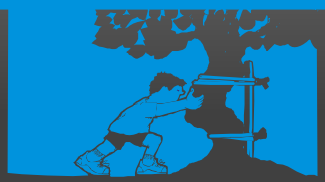
UKBB
Universitäts-Kinderspital beider Basel

Clinical Presentation & Evaluation and Staging

Pediatric Orthopedic Department
University of Basel, Switzerland

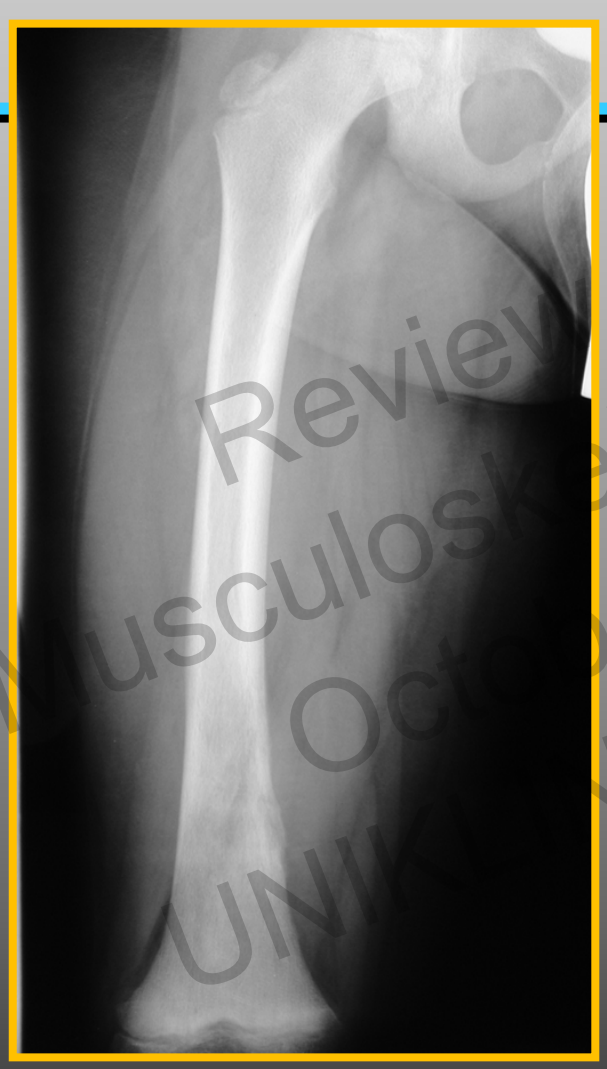


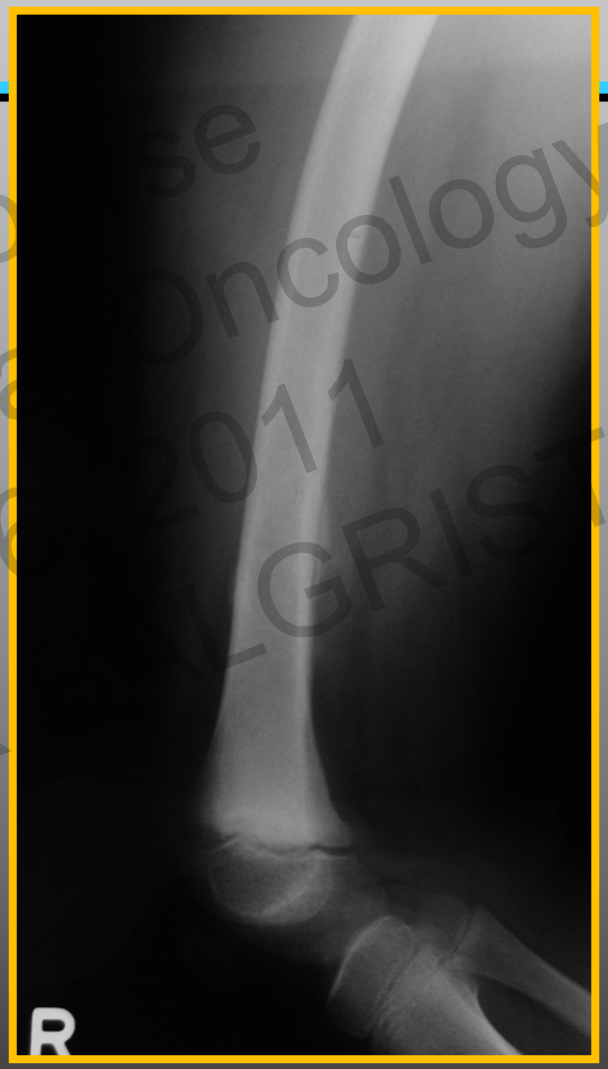
- 8-year old boy
- for 10 days pain in his right knee and thigh

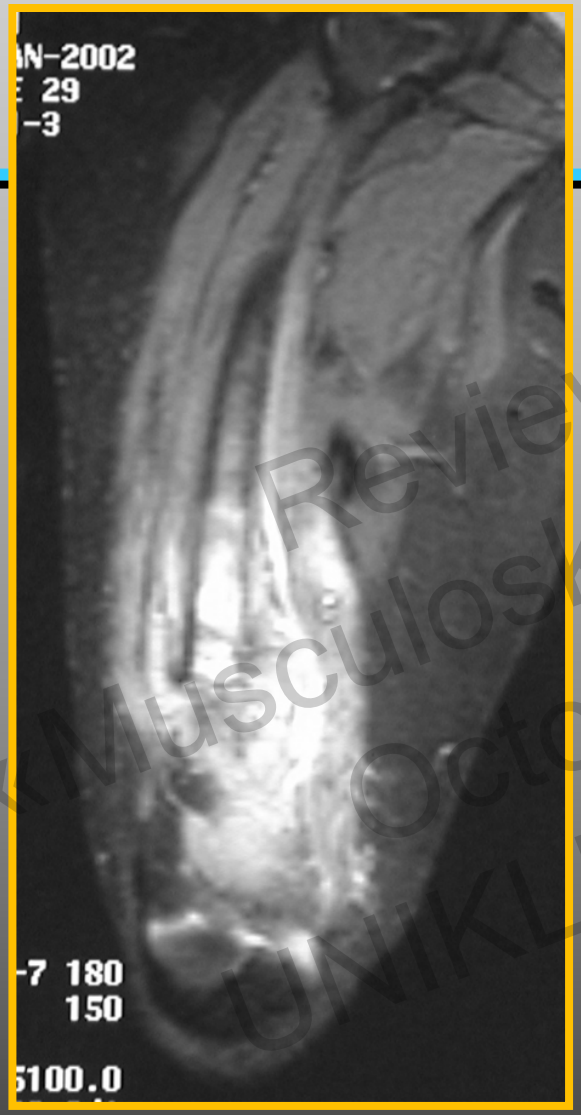
- *questions?*
- *measures to take?*

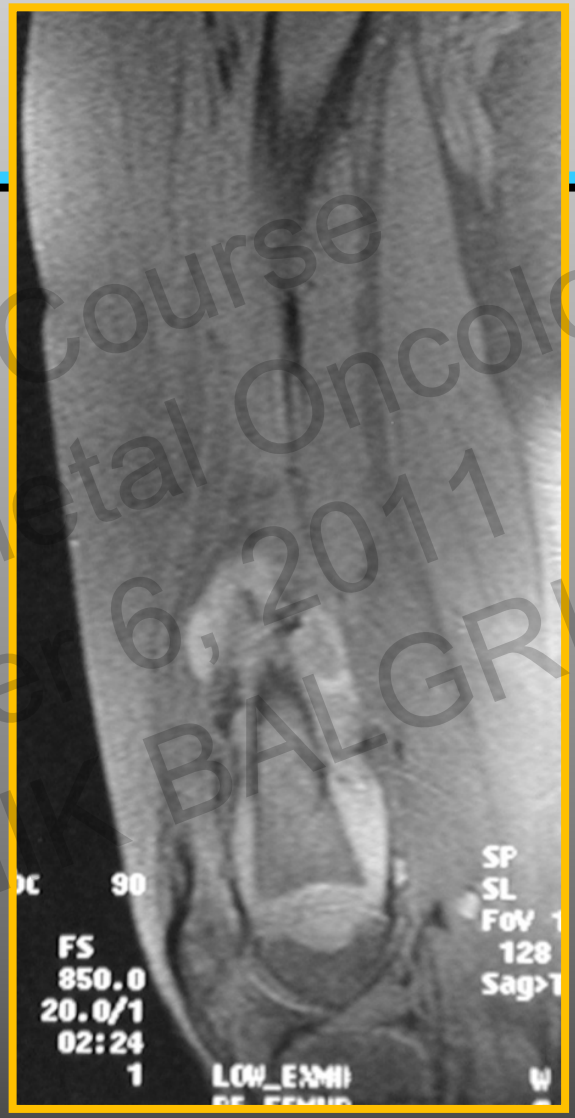
- pain persists now for 4 weeks
- they still are about the same

- *questions?*
- *measures to take?*

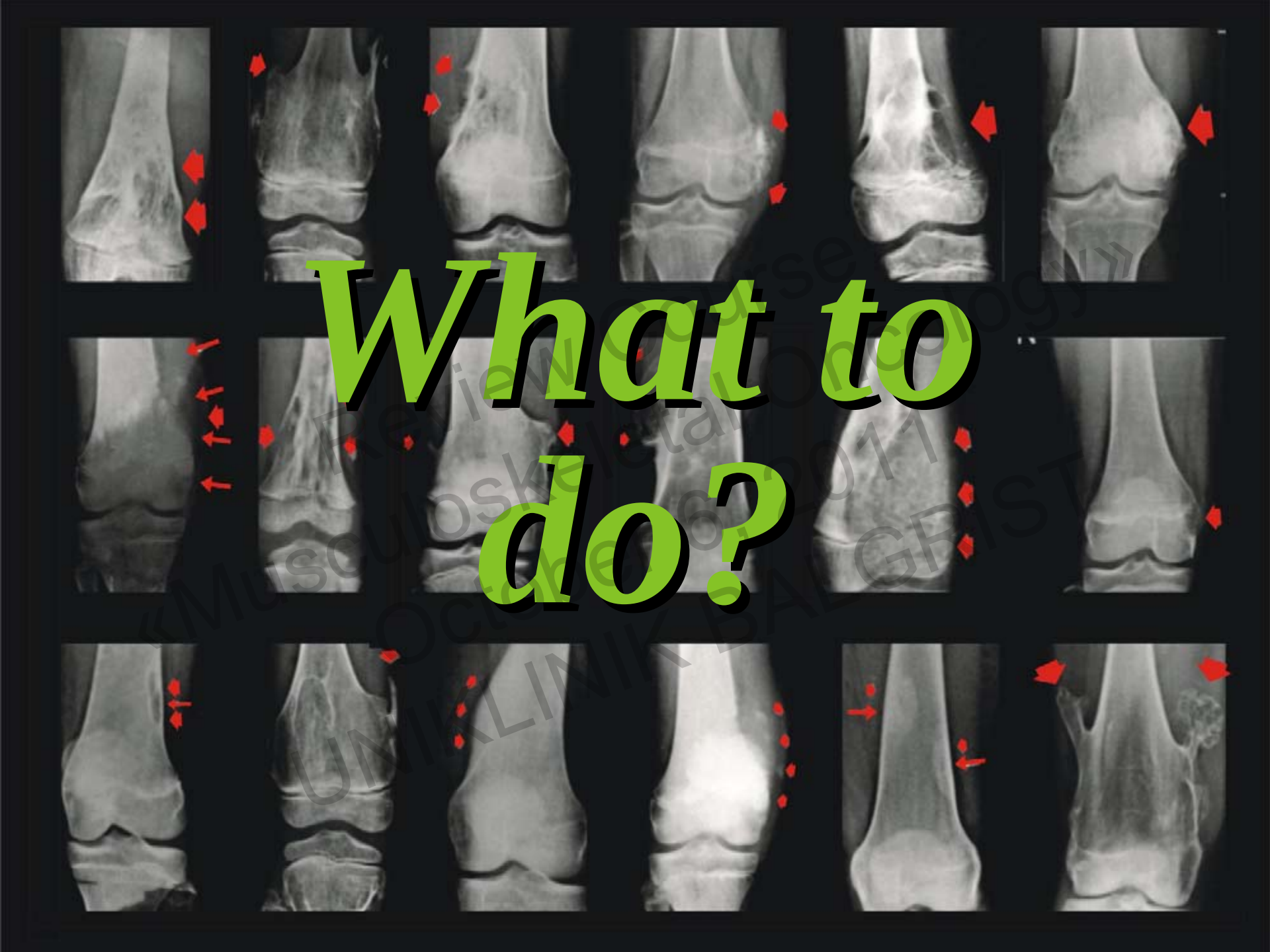








S.I., m., 8+2


What to do?

Evaluation and Staging

- history
- clinical findings
- laboratory findings
- imaging
- biopsy
- staging

History

- pain?
- functional limitations?
- weight?

Pain history

when did it start?

trauma?

activity?

night pain?

duration?

how often?

Night pain

- high-grade malignant bone tumors (osteosarcomas, Ewing-sarcomas etc.)
- bone producing benign bone tumors (osteoid-osteoma, osteoblastoma)
- tumors with instability because of (micro-)fractures (mainly metastases)
- infections (bone and soft-tissue)
- growing pain (usually bilateral)

No night pain in

- **all soft tissue sarcomas** (incl. high-grade sarcomas)
- **all benign soft tissue tumors** (except for mechanical pain)
- **low grade malignant bone tumors**
- **benign bone tumors** (except bone producing tumors)

Clinical findings

- consistency
- displaceability of the skin
- displaceability on the bone
- tenderness
- reddening
- warming up

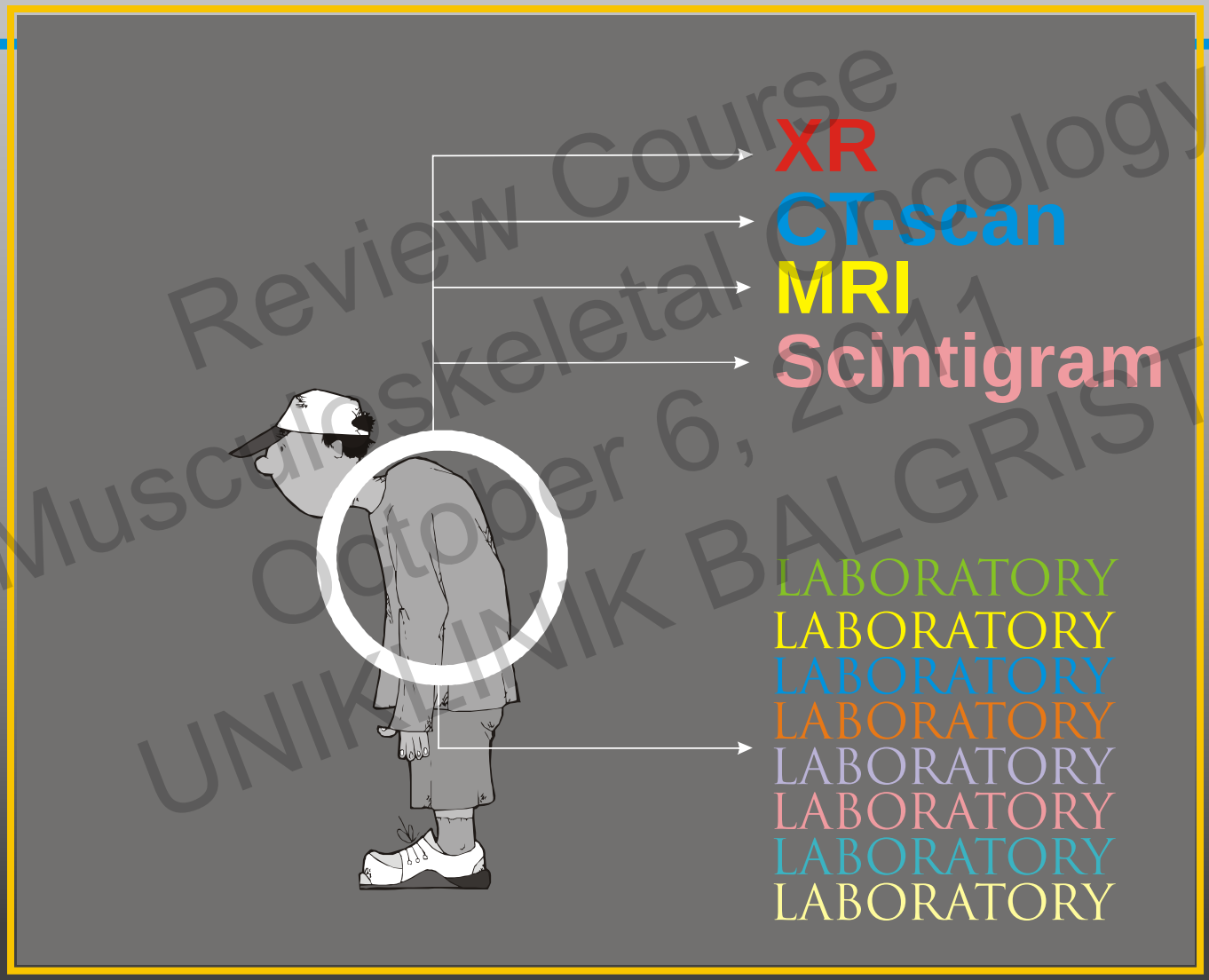
Functional limitations

- limping
- reduced ROM
- reduced force

Laboratory findings

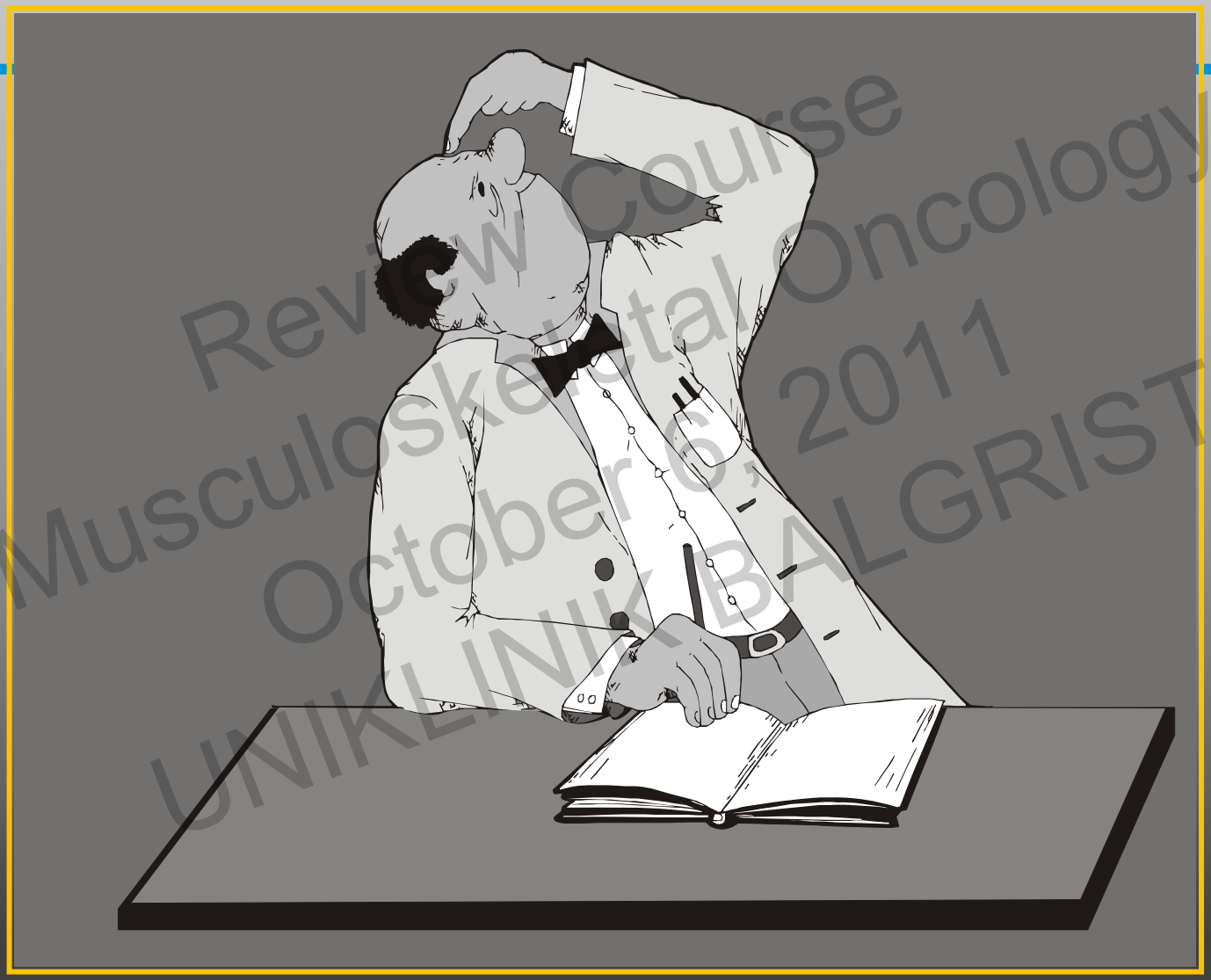
- **serum alkaline phosphatase (osteosarcoma?)**
- **lactate dehydrogenase (LDH) (Ewing‘s sarcoma?)**
- **sedimentation rate (Ewing‘s sarcoma)**
- **catecholamines (neuroblastoma?)**
- **monoclonal proteins (plasmocytoma?)**

Making the diagnosis (I)



Making the diagnosis (II)

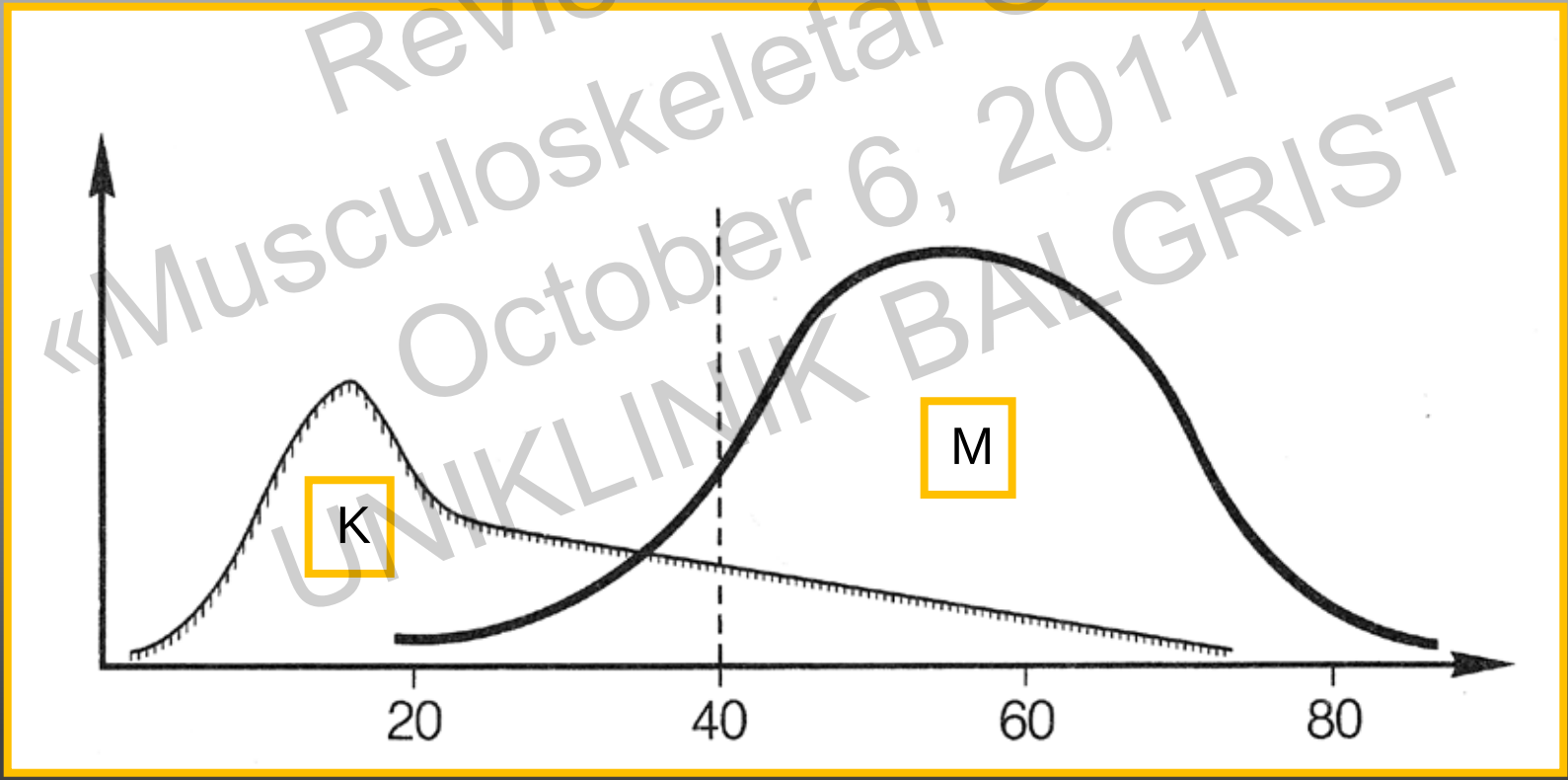
Making the diagnosis (III)



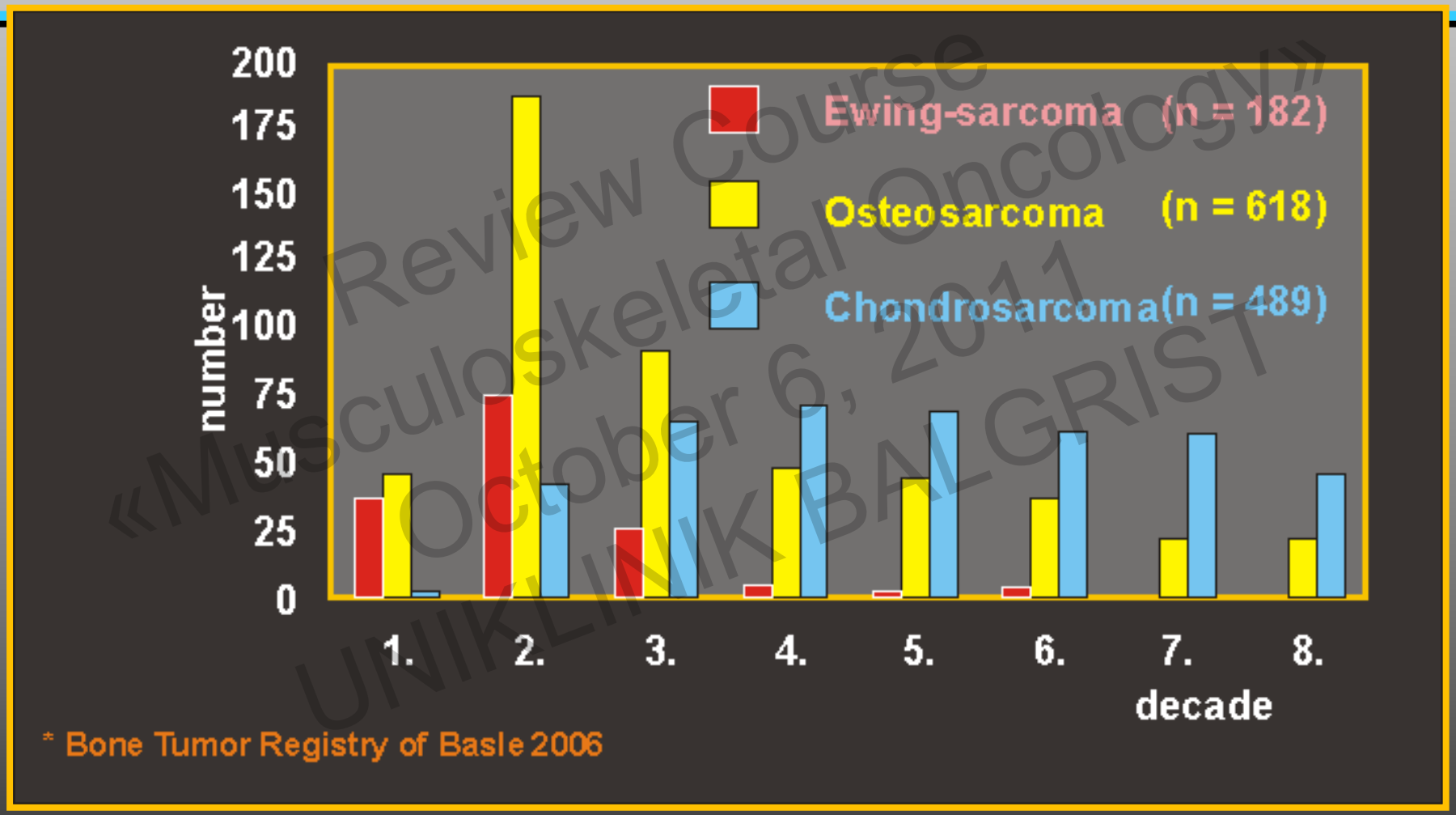
Incidence of Bone Tumors

~0.5 % of all malignant tumors: Bone tumors
(without plasmocytoma: ~0.5%)

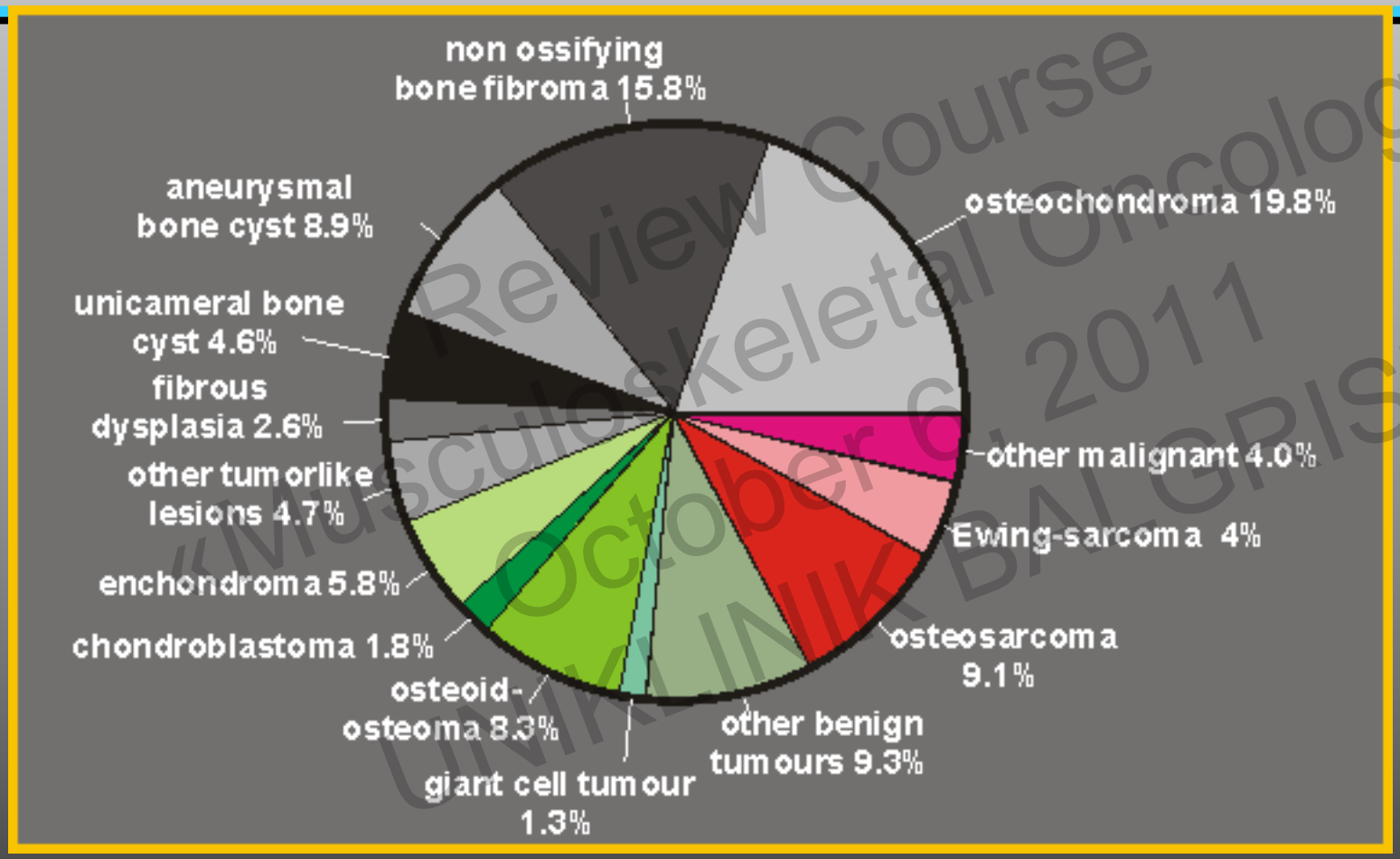
Metastases (CUP) ~ 6-8x more frequent



Age at diagnosis of the 3 most common malignant bone tumors



Bone tumors in children and adolescents



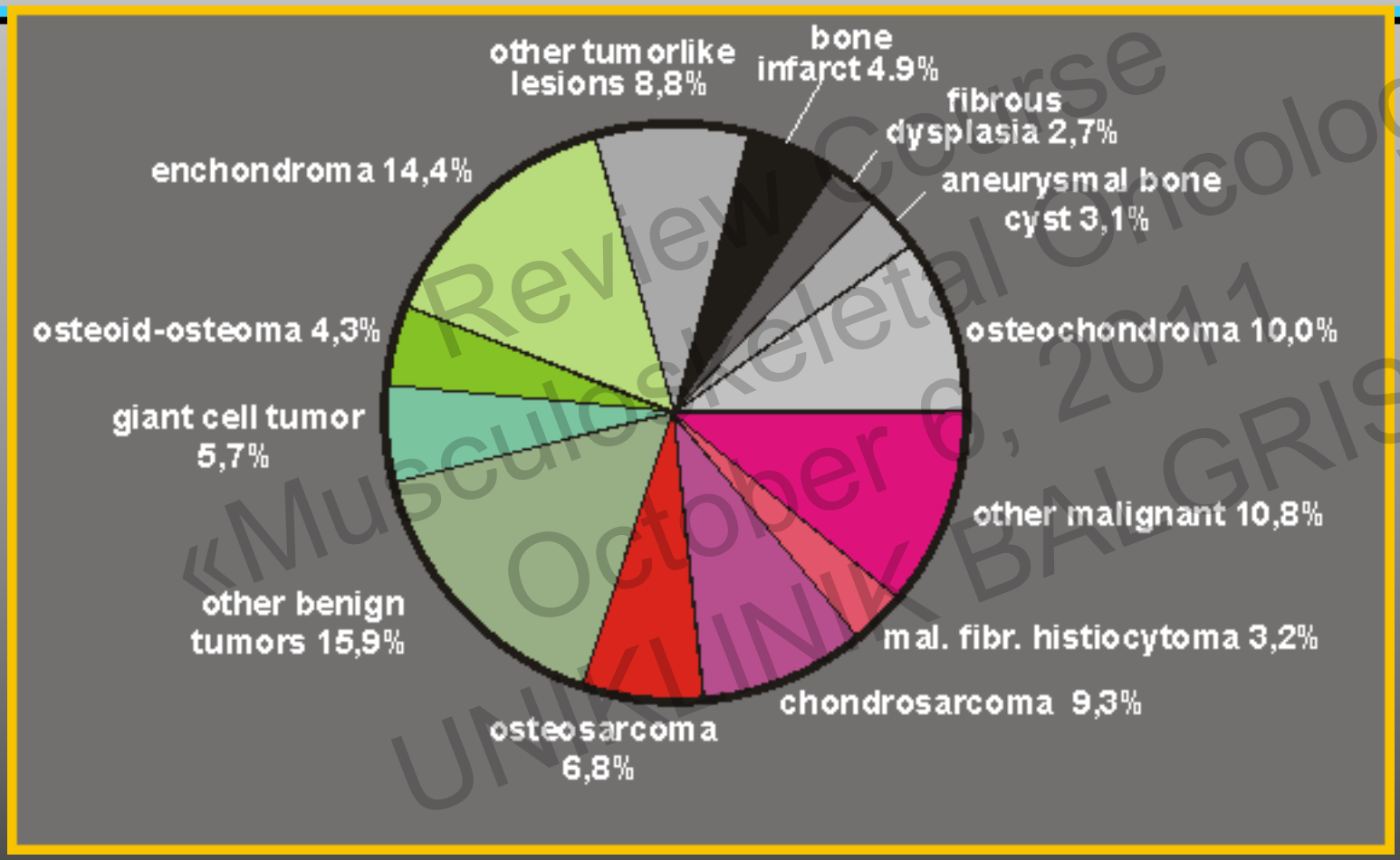
Benign Tumors

Malignant Tumors

Tumorlike Lesions

Basle Bone Tumour Registry, tumors at age < 20 y., n=4408

Bone tumors in adults



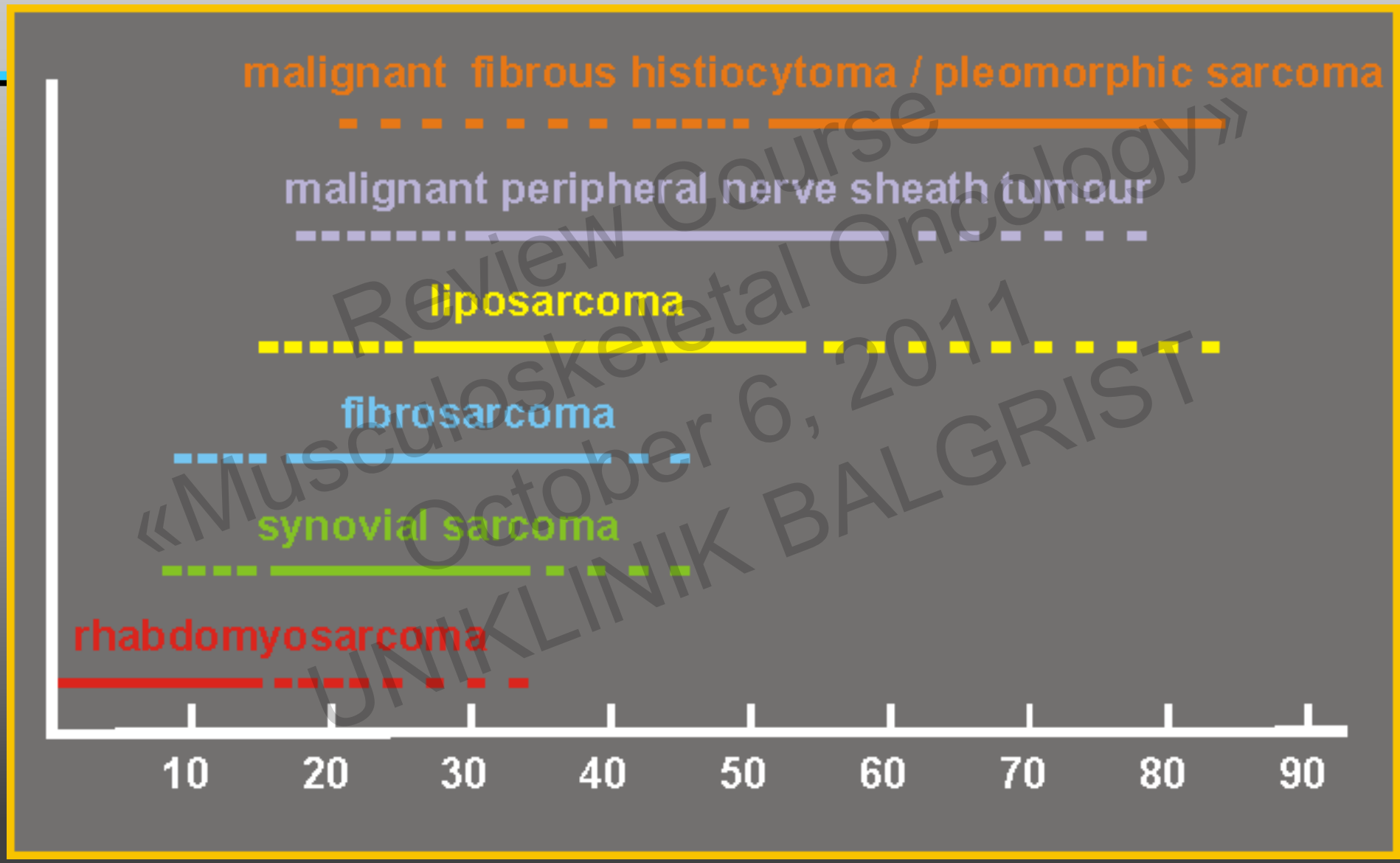
Benign Tumors

Malignant Tumors

Tumorlike Lesions

Basle Bone Tumour Registry, tumors at age > 20 y., n=7534

Age related occurrence of soft-tissue tumors

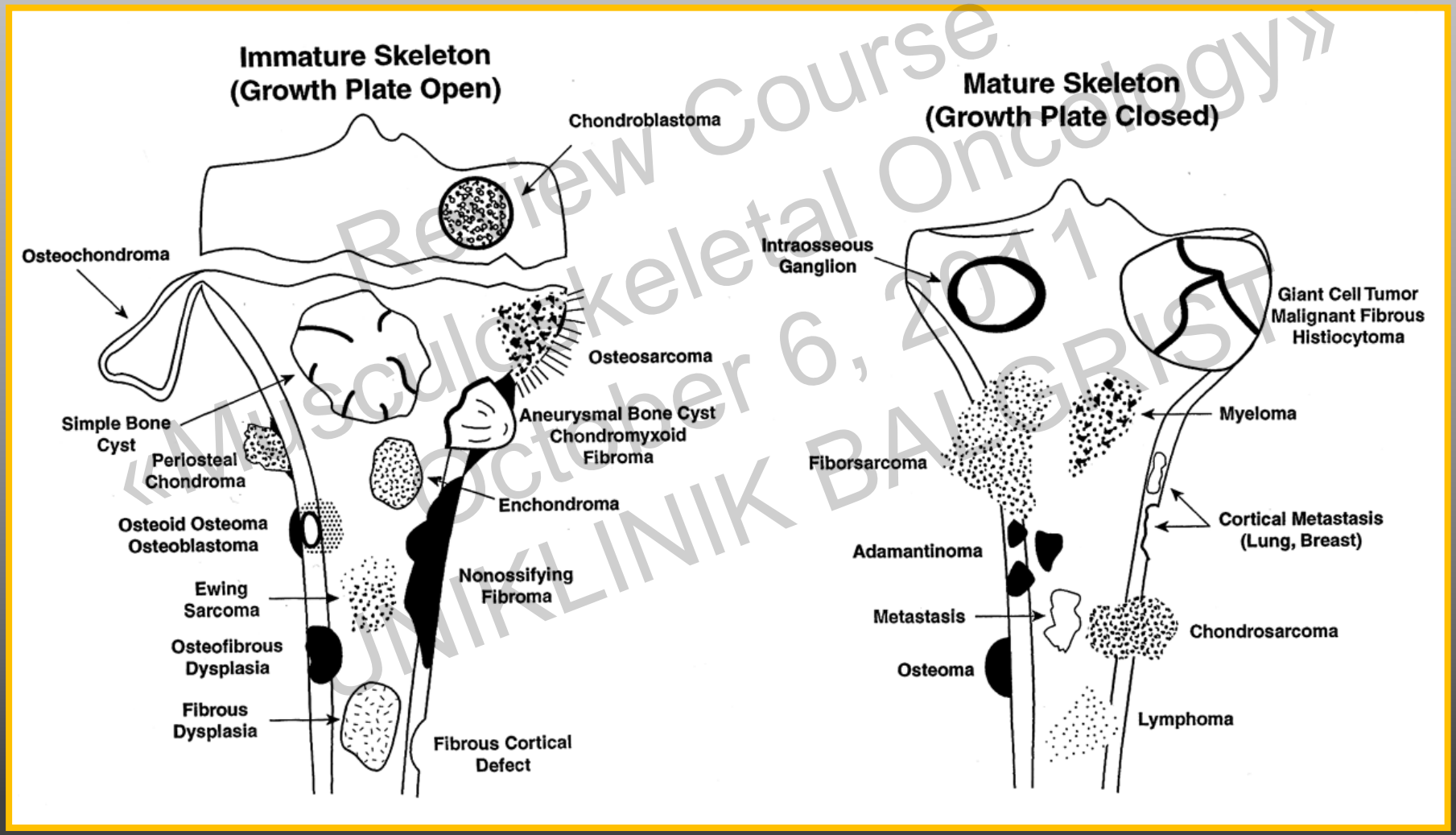


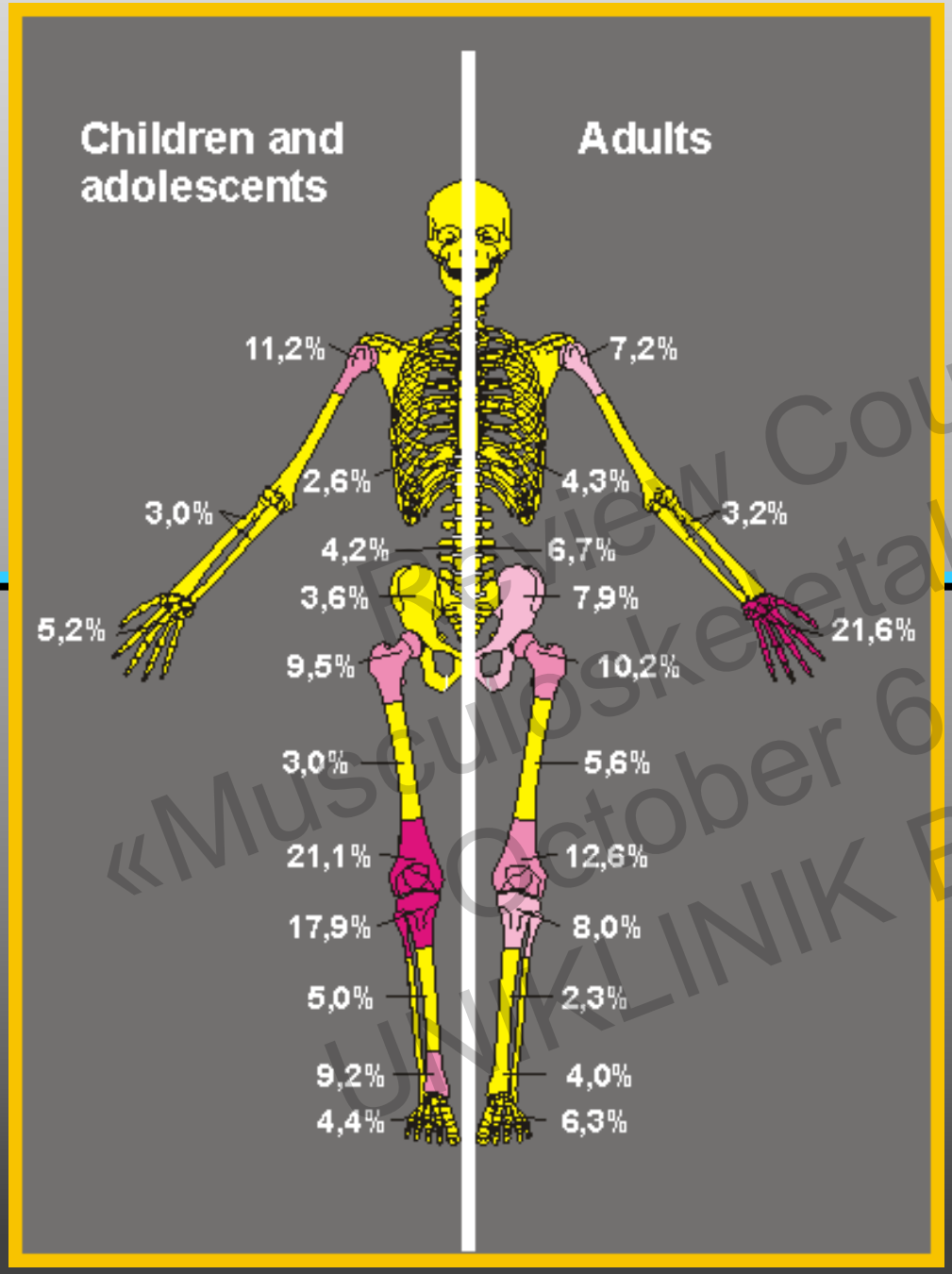
Topography within the bone

benign vs malignant

epiphysis	chondroblastoma, giant cell tumors clear cell chondrosarcoma,
metaphysis	non-ossifying fibroma, solitary bone cyst, aneurysmal bone cyst, giant cell tumors, osteoblastoma, chondroma, osteosarcoma, (chondrosarcoma)
diaphysis	fibrous dysplasia, osteofibrous dysplasia (Campanacci) Ewing-sarcoma, primitive neuroectodermal tumor (PNET), Non-Hodgkin-lymphoma (NHL) of bone, adamantinoma, chondrosarcoma

Age distribution and topography of bone tumors





Localization of bone tumors in children and adults

Screening for metastases

- x-ray of thorax, when suspicious CT-scan
- ultrasound abdomen, when suspicious CT-scan
- PET (or PET-CT)
- occasionally a whole body MRI is more efficient

Classification of Bone Tumors

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Benign Tumors

Malignant Tumors

Tumor-like Lesions

Classification of bone tumors

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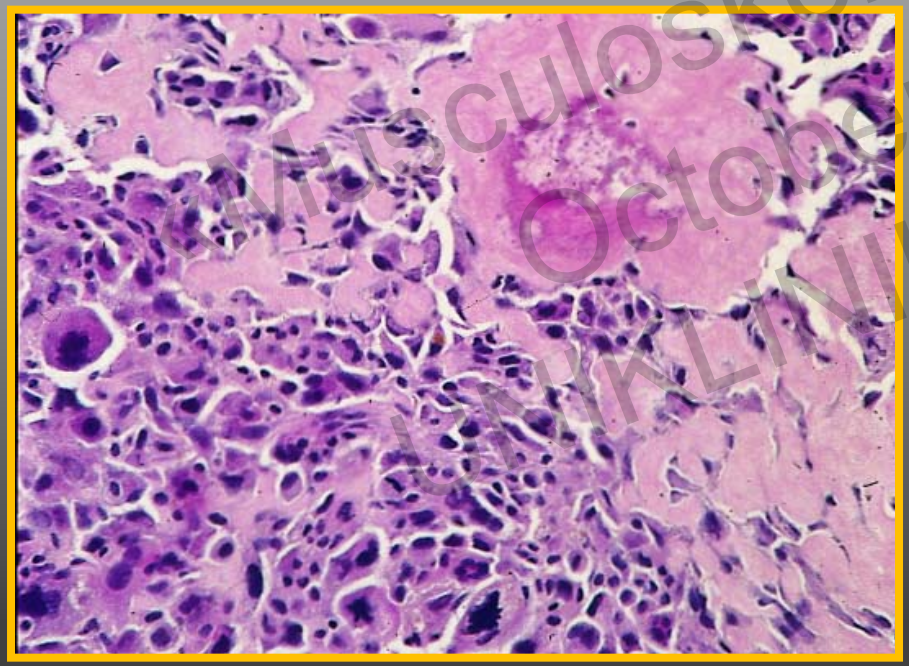
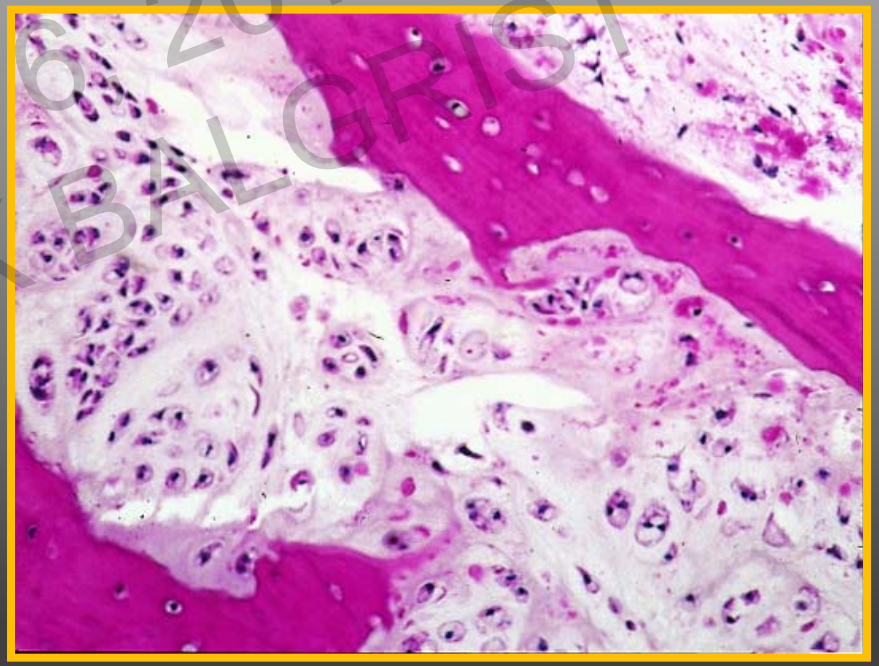
Histological Classification(WHO):

accoding to the type of tissue imitated by the tumor cells

bone > Osteosarcoma **cartilage > Chondrosarcoma**





Classification of Bone Tumors

- Tumors that produce bone
- Tumors that produce cartilage
- Tumors containing giant cells
- Tumors of the bone marrow
- Tumors that produce vessels
- Tumors that produce connective tisssue
- Other tumors
- Tumor-like lesions

Classification of Bone Tumors

benign vs malignant

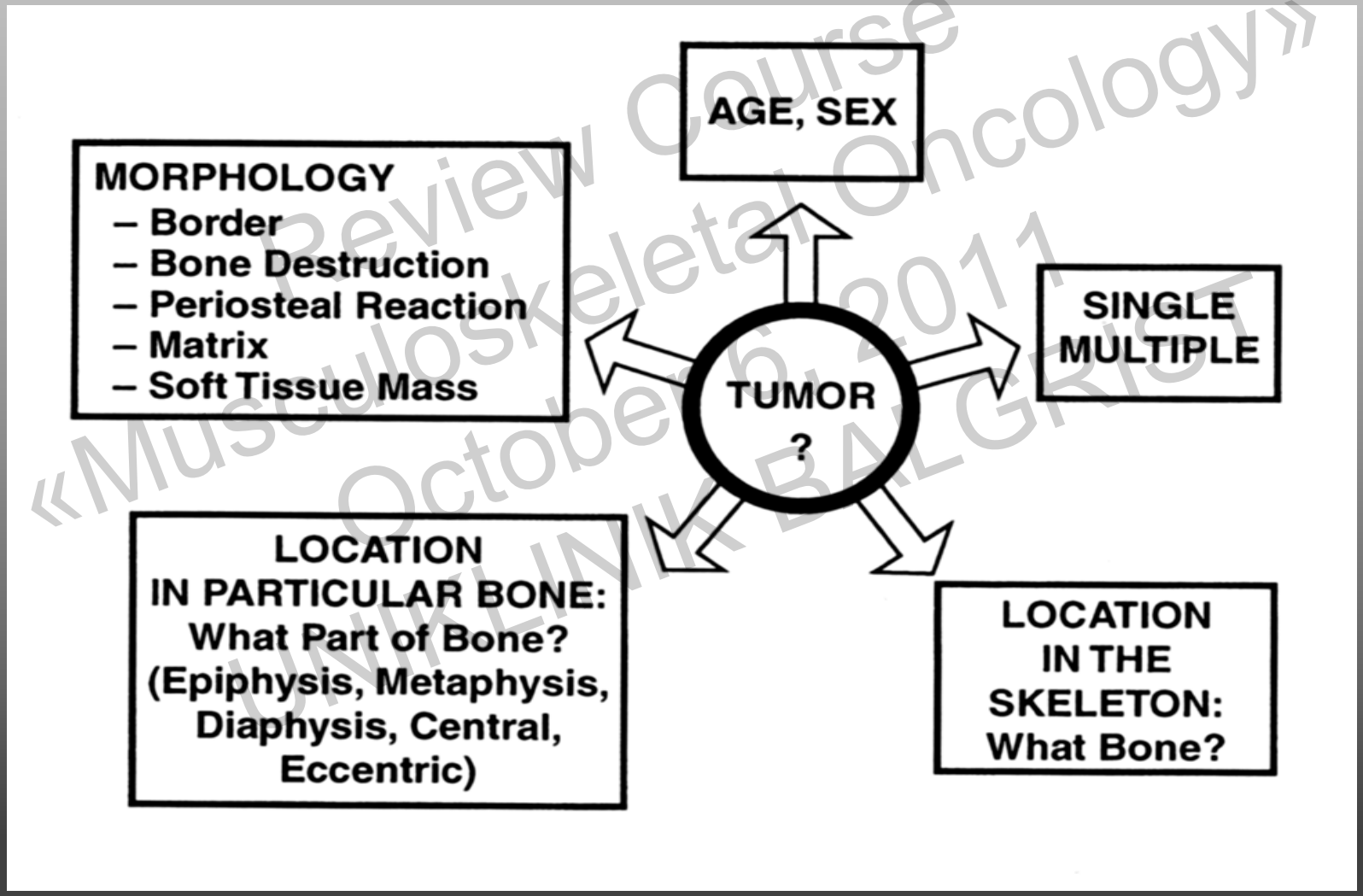
Osteogenic tumors	osteoma, osteoid-osteoma, osteblastoma osteosarcoma, parosteal osteosarcoma, low-grade central osteosarcoma
cartilage producing tumors	enchondroma, osteochondroma, periosteal chondroma, chondroblastoma chondrosarcoma, clear cell chondrosarcoma, dedifferentiated chondrosarcoma
fibrous tissue producing tumors	benign fibrous histiocytoma, desmoplastic fibroma malignant-fibrous histiocytoma, fibrosarcoma

Classification of Bone Tumors

benign vs malignant

tumors containing giant cells	giant cell tumors
tumor producing vessels	hemingioma, hemangioendothelioma angiosarcoma
tumors of the bone marrow	Ewing-sarcoma, primitive neuroectodermal tumor (PNET), Non-Hodgkin-lymphoma (NHL) of bone
other tumors	chordoma, adamantinoma
tumor-like lesions	solitary bone cyst, aneurysmal bone cyst, Langehans'cell histiocytosis, fibrous dysplasia, non-ossifying fibroma

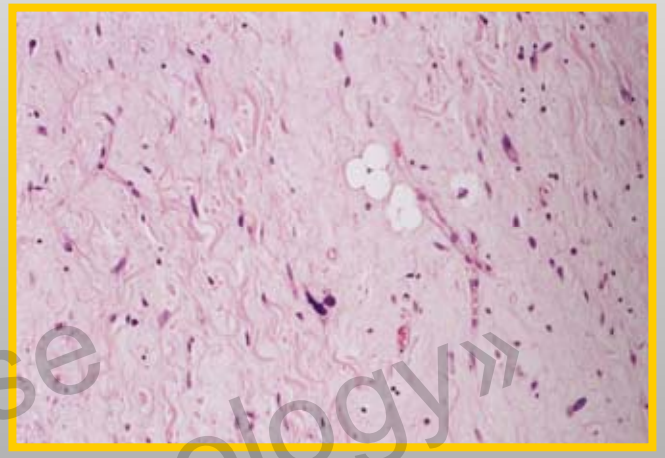
Diagnostic procedure in bone tumors



Exam-ples of histo-logical grading

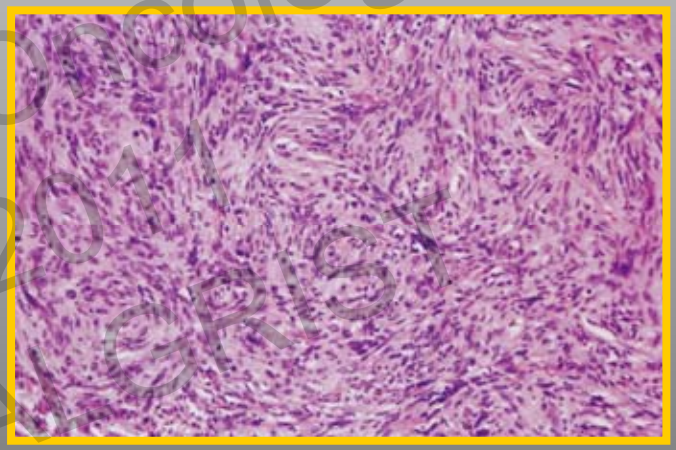
Grade 1

liposarcoma, well differentiated



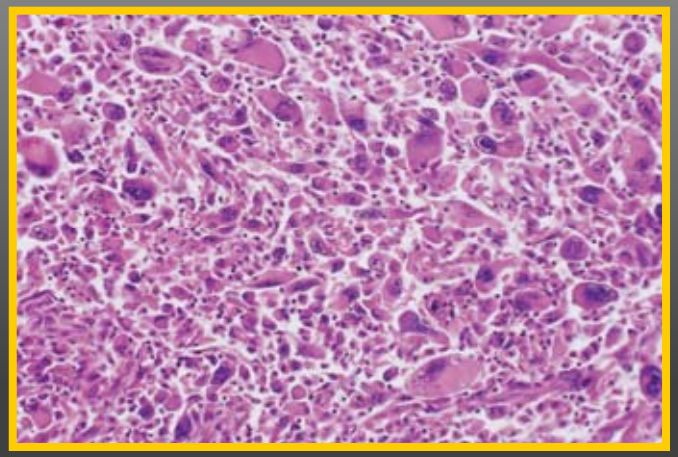
Grade 2

malignant peripheral nerve sheath tumor (MPNST)

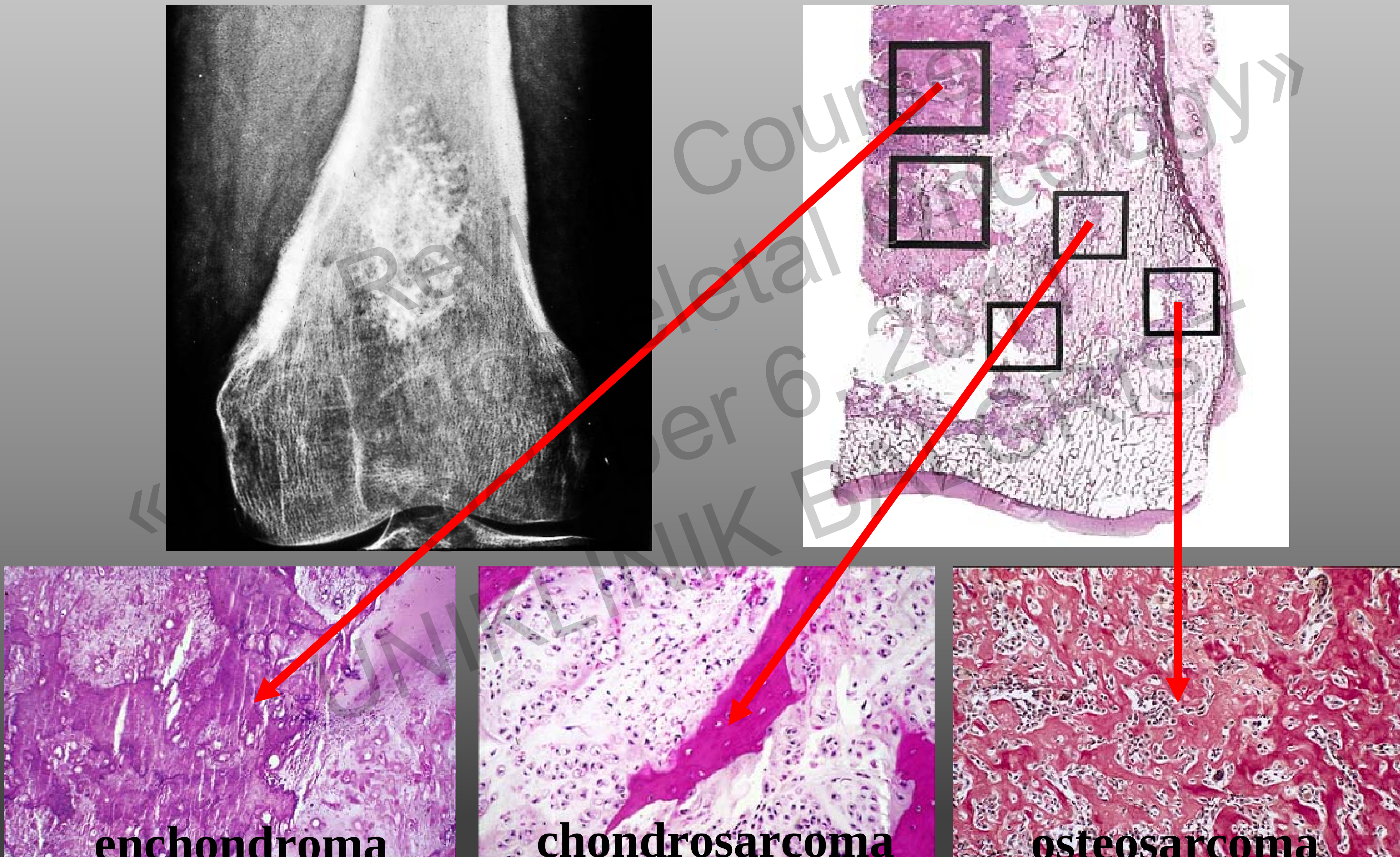


Grade 3

pleomorphic rhabdomyosarcoma



Dedifferentiated chondrosarcoma



Staging-system according to Enneking

G-Grade	histolocial differentiation
T=Site	anatomical localisation, compartments
M=Metastases	

Staging-system according to Enneking

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Stage:	Grade:	Site:	Metastases
IA	G1 (diff.)	T1 (intracomp.)	M0 (none)
IB	G1 (diff.)	T2 (extracomp.)	M0 (none)
IIA	G2 (dediff.)	T1 (intracomp.)	M0 (none)
IIB	G2 (dediff.)	T2 (extracomp.)	M0 (none)
IIIA	G2 (dediff.)	T1 (intracomp.)	M1 (present)
IIIB	G2 (dediff.)	T2 (extracomp.)	M1 (present)

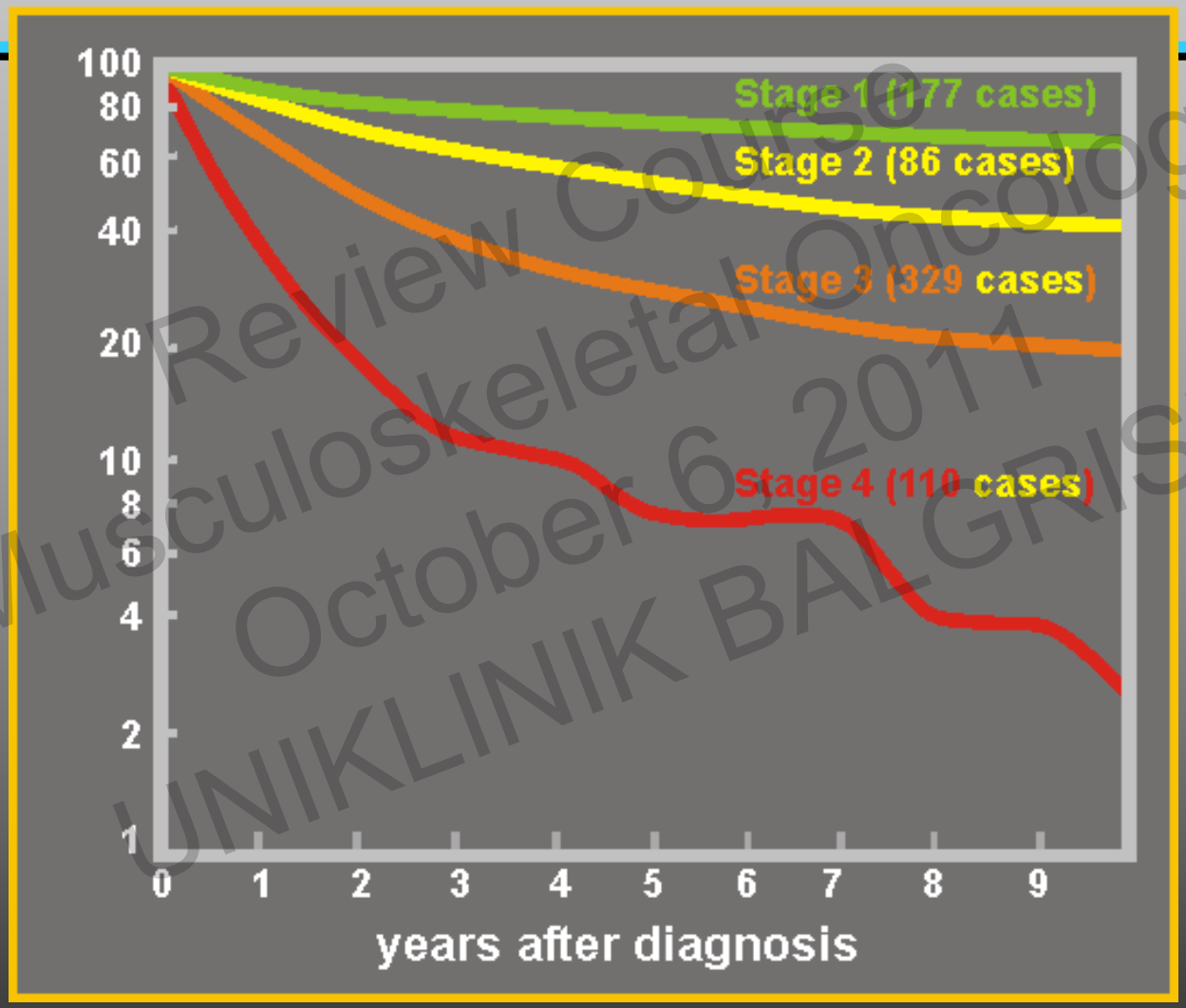
American Joint Committee (AJC) staging system of soft tissue tumors

G-Grade	histolocial differentiation
T=Site	anatomical localisation, compartments
N=regional lymph nodes	
M=distant metastases	

The AJC Staging-system of soft-tissue tumors

Stage:	Grade:	Site:	Lymph-nodes	Metastases
IA	G1 (diff.)	T1 (intracomp.)	N0 (none)	M0 (none)
IB	G1 (diff.)	T2 (extracomp.)	N0 (none)	M0 (none)
IIA	G2 (moder.)	T1 (intracomp.)	N0 (none)	M0 (none)
IIB	G2 (moder.)	T2 (extracomp.)	N0 (none)	M0 (none)
IIIA	G3,4 (dediff.)	T1 (intracomp.)	N0 (none	M0 (none)
IIIB	G3,4 (dediff.)	T2 (extracomp.)	N0 (none)	M0 (none)
IVA	G1-4	T1-2	N1 (present)	M0 (none)
IVB	G1-4	T1-2	N0 (+/-)	M1 (present)

Survival rates of soft tissue tumors depending on stage



Assessment of bone tumors

Heterogenous structure of bone tumors

Histological-radiological-clinical correlation is always necessary

Bone tumors
are
interdisciplinary problems

Multidisciplinary Tumor Center in Basel

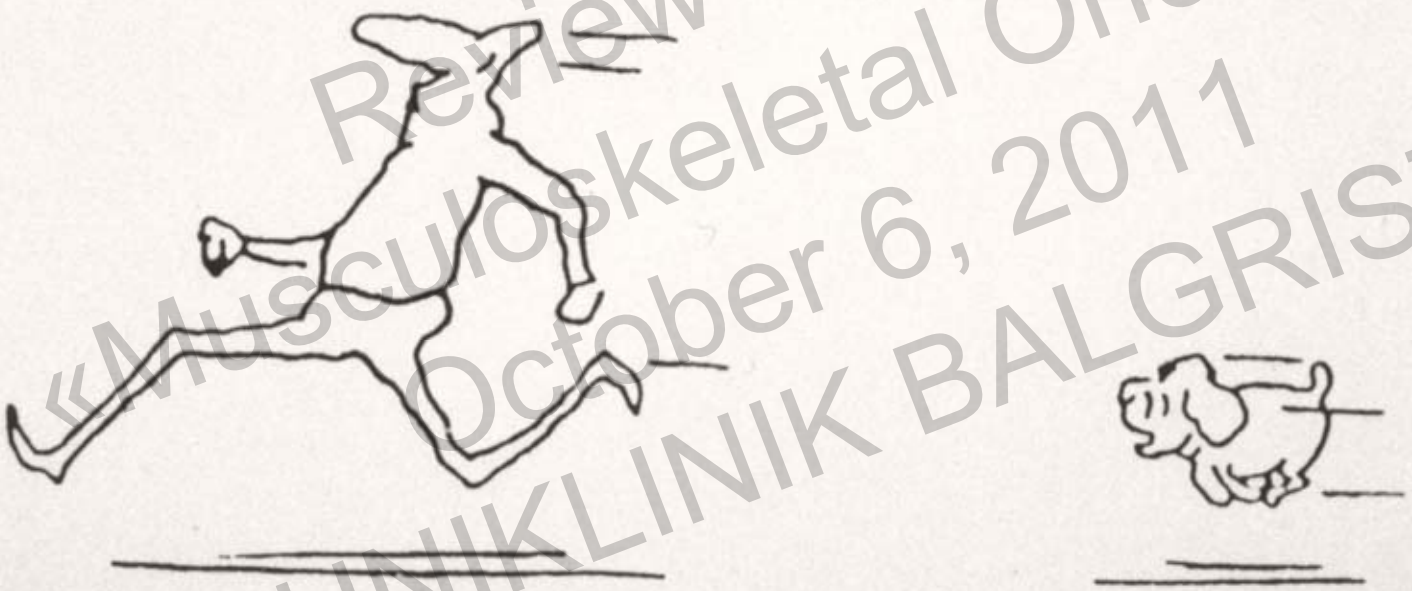
Universitäts-Kinderspital
beider Basel (UKBB)

Universitätsspital Basel (USB)

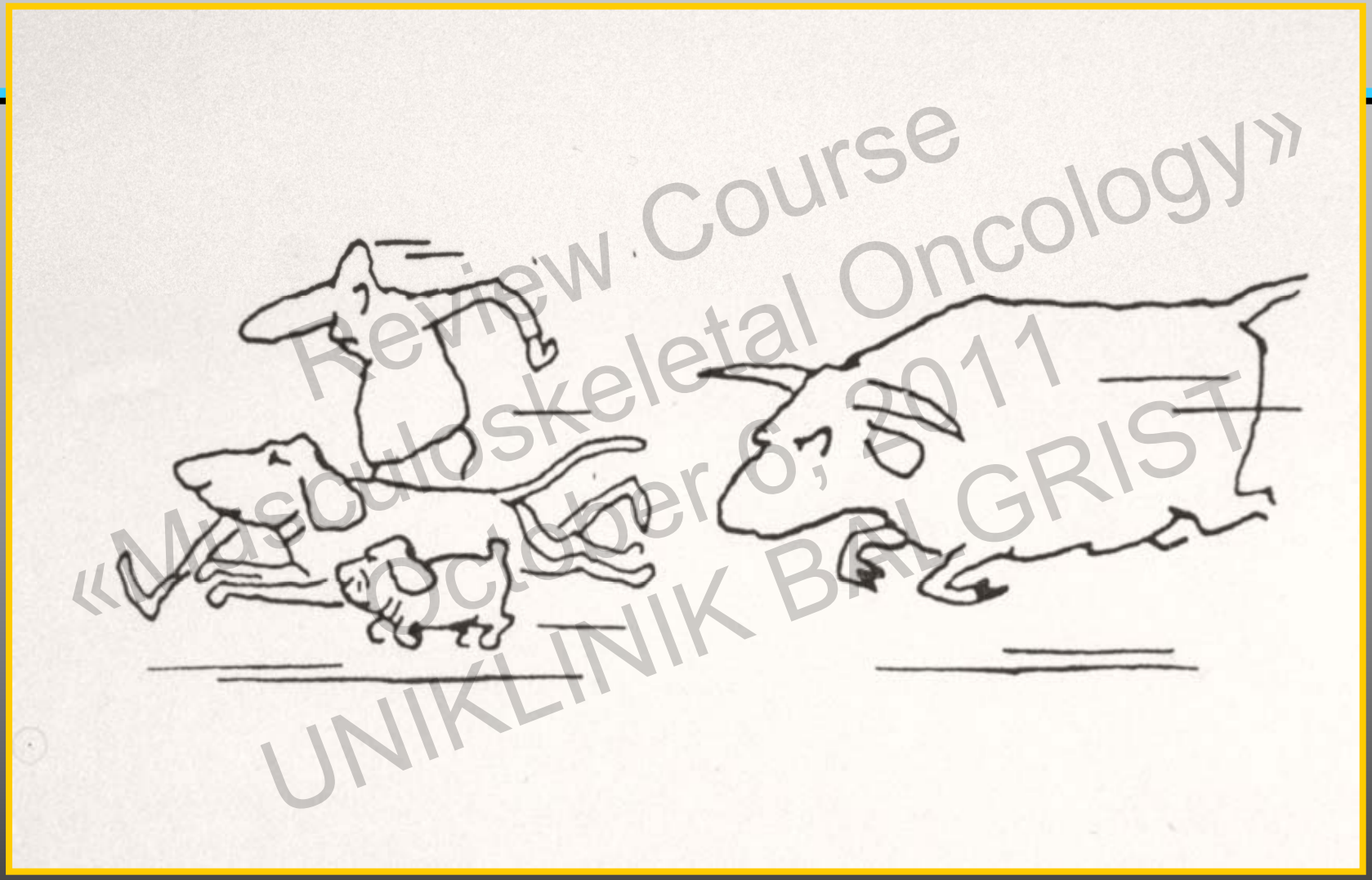
Interdisziplinäres Knochen- und Weichteiltumorzentrum der Universität Basel (KWUB)'

Leitung
Prof. Dr. F. Hefti, Kinderorthopädie, Tumorchirurgie (UKBB)

Mitglieder
Prof. Dr. C. Kettelhack, Viszeral-, Tumorchirurgie (USB)
Dr. A. Krieg, Kinderorthopädie, Tumorchirurgie (UKBB)
PD Dr. M. Haug, Plastische Chirurgie (USB)
Prof. Dr. T. Kühne, Kinderonkologie (UKBB)
Fr. Dr. F. Krasniqi, Onkologie (USB)
PD Dr. M. Gross, Radioonkologie (USB)
Prof. Dr. G. Jundt, Pathologie (USB)
Fr. PD Dr. E. Bruder, Pathologie (USB)
Prof. Dr. A. Nidecker, Radiologie
Dr. J. Schneider, Radiologie (UKBB)
Dr. U. Studler, Radiologie (USB)
Dr. T. Wischer, Radiologie



Review Course
«Musculoskeletal Oncology»
October 6, 2011
UNIKLINIK BALGRIST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