Epidemiology of Bone and Soft Tissue Sarcomas (STS)

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TABLE 1–3 HISTOLOGIC CLASSIFICATION OF SOFT TISSUE TUMORS

- Fibrous tumors
  - Benign
    - Nodular fasciitis (including intravascular and cranial types)
    - Proliferative fasciitis and myositis
    - Organ-associated pseudosarcomatous myofibroblastic proliferations
    - Ischemic fasciitis (atypical decubital fibroplasia)
    - Fibroma (dermal, tendon sheath, nuchal types)
    - Elastofibroma
    - Nasopharyngeal angiofibroma
    - Giant cell angiofibroma
    - Keloid
    - Desmoplastic fibroblastoma (collagenous fibroma)
    - Fibrous hamartoma of infancy
    - Infantile digital fibromatosis
    - Myofibroma and myofibromatosis
    - Hyalin fibromatosis
    - Gingival fibromatosis
    - Fibromatosis colli
    - Calcifying aponeurotic fibroma
    - Calcifying fibrous pseudotumor
    - Infantile-type fibromatosis
  - Intermediate
    - Adult-type fibromatosis
      - Superficial (including palmar, plantar, penile fibromatosis, knuckle pads)
      - Deep (including extraabominal, abdominal, intraabdominal, mesenteric, pelvic fibromatosis)
    - Inflammatory myofibroblastic tumor (inflammatory fibrosarcoma)
    - Infantile fibrosarcoma
  - Malignant
    - Adult-type fibrosarcoma
      - Usual type
      - Myxoid type (myxofibrosarcoma, low grade myxoid malignant fibrous histiocytoma)
      - Low grade fibromyxoid type with or without rosettes (low grade fibromyxoid sarcoma)
      - Sclerosing epithelioid type
- Fibrohistiocytic tumors
  - Benign
    - Fibrous histiocytoma (cutaneous and deep)
      - Cellular
      - Epithelioid
    - Juvenile xanthogranuloma
    - Reticulohistiocytoma
    - Xanthoma
    - Extranodal (soft tissue) Rosai-Dorfman disease
  - Intermediate
    - Atypical fibroxanthoma
    - Dermatofibrosarcoma protuberans (including pigmented forms)
    - Giant cell fibroblastoma
    - Angiomatoid fibrous histiocytoma
    - Plexiform fibrohistiocytic tumor
    - Soft tissue giant cell tumor of low malignant potential
  - Malignant
    - Malignant fibrous histiocytoma
      - Storiform-pleomorphic type
      - Myxoid type
      - Giant cell type (malignant giant cell tumor of soft parts)
      - Inflammatory type
- Lipomatous tumors
  - Benign
    - Lipoma [solitary, multiple, cutaneous, deep (including intramuscular and perineural)]
    - Angiolipoma
    - Myolipoma
    - Chondroid lipoma
    - Spindle cell/pleomorphic lipoma
    - Angiomyolipoma
    - Myelolipoma
    - Hibernoma
    - Lipoblastoma or lipoblastomatosis
    - Lipomatosis
      - Diffuse lipomatosis
      - Cervical symmetric lipomatosis (Madelung's disease)
      - Pelvic lipomatosis
  - Intermediate
    - Atypical lipoma (well differentiated liposarcoma of superficial soft tissue, atypical lipomatous tumor)
  - Malignant
    - Well differentiated liposarcoma
      - Lipoma-like
      - Sclerosing
      - Inflammatory
      - Spindle cell
    - Myxoid-round cell liposarcoma
    - Pleomorphic liposarcoma
    - Dedifferentiated liposarcoma
- Smooth muscle tumors and related lesions
  - Benign
    - Leiomyoma
    - Angiomyoma
    - Angiomyofibroblastoma
    - Palisaded myofibroblastoma of lymph node
    - Intravenous leiomyomatosis
    - Leiomyomatosis peritonealis disseminata
  - Malignant
    - Leiomyosarcoma
- Extragastrointestinal (soft tissue) stromal tumors
  - Benign
    - Benign extragastrointestinal stromal tumor
    - Benign extragastrointestinal autonomic tumor
  - Malignant
    - Malignant extragastrointestinal stromal tumor
    - Malignant extragastrointestinal autonomic nerve tumor
- Skeletal muscle tumors
  - Benign
    - Cardiac rhabdomyoma
    - Adult rhabdomyoma
    - Fetal rhabdomyoma
      - Myxoid (classic)
      - Intermediate (cellular, juvenile)
  - Malignant
    - Embryonal rhabdomyosarcoma
      - Usual type
      - Botryoid type
      - Spindle cell type
    - Alveolar rhabdomyosarcoma
    - Pleomorphic rhabdomyosarcoma
    - Rhabdomyosarcoma with ganglion cells (ectomesenchymoma)
- Tumors of blood and lymph vessels
  - Benign
    - Papillary endothelial hyperplasia
    - Hemangioma
      - Capillary hemangioma (including juvenile)
      - Cavernous hemangioma (including sinusoidal)
      - Venous hemangioma
      - Epithelioid hemangioma (angiolymphoid hyperplasia)
      - Pyogenic granuloma
      - Acquired tufted hemangioma
      - Hobnail hemangioma
      - Spindle cell hemangioma
    - Lymphangioma
    - Lymphangiomyoma and lymphangiomyomatosis
    - Angiomatosis
    - Lymphangiomatosis
  - Intermediate
    - Epithelioid hemangioendothelioma
    - Hobnail hemangioendothelioma
      - Retiform type (retiform hemangioendothelioma)
      - Dabska type (endovascular papillary angioendothelioma)
    - Kaposiform hemangioendothelioma
  - Malignant
    - Angiosarcoma
    - Kaposi's sarcoma
- Perivascular tumors
  - Benign
    - Glomus tumor
      - Usual type
      - Glomangioma
      - Glomangiomyoma
      - Glomangiomatosis
    - Benign hemangiopericytoma/solitary fibrous tumor
    - Myopericytoma
  - Malignant
    - Malignant glomus tumor
    - Malignant hemangiopericytoma/malignant solitary fibrous tumor
- Synovial tumors
  - Benign
    - Tenosynovial giant cell tumor
      - Localized type
      - Diffuse type
  - Malignant
    - Malignant tenosynovial giant cell tumor
- Mesothelial tumors
  - Benign
    - Adenomatoid tumor
  - Intermediate
    - Multicystic mesothelioma
    - Well differentiated papillary mesothelioma
  - Malignant
    - Diffuse mesothelioma
      - Epithelial type
      - Sarcomatoid type
      - Biphasic type
- Peripheral nerve sheath tumors and related lesions
  - Benign
    - Traumatic neuroma
    - Glial heterotopia
    - Mucosal neuroma
    - Pacinian neuroma
    - Palisaded encapsulated neuroma
    - Morton's interdigital neuroma
    - Nerve sheath ganglion
    - Neuromuscular hamartoma
    - Neurofibroma and neurofibromatosis
      - Usual type (localized)
      - Diffuse
      - Plexiform
      - Epithelioid
    - Schwannoma and schwannomatosis
      - Usual type
      - Cellular schwannoma
      - Plexiform schwannoma
      - Degenerated (ancient) schwannoma
      - Epithelioid schwannoma
      - Neuroblastoma-like schwannoma
    - Melanotic schwannoma
    - Perineurioma
      - Intraneural perineurioma (localized hypertrophic neuropathy)
      - Extraneural (soft tissue) perineurioma
    - Granular cell tumor
    - Neurothekeoma
    - Ectopic meningioma
  - Malignant
    - Malignant peripheral nerve sheath tumor (MPNST)
      - Usual type
      - MPNST with rhabdomyoblastic differentiation (malignant Triton tumor)
      - Glandular malignant schwannoma
      - Epithelioid MPNST
      - MPNST arising in a schwannoma
      - MPNST arising in a ganglioneuroma
    - Malignant granular cell tumor
    - Clear cell sarcoma of the tendon and aponeurosis
    - Malignant melanocytic schwannoma
    - Ectopic ependymoma
- Primitive neuroectodermal tumors and related lesions
  - Benign
    - Ganglioneuroma
    - Pigmented neuroectodermal tumor of infancy (retinal anlage tumor)
  - Malignant
    - Neuroblastoma
    - Ganglioneuroblastoma
    - Ewing's sarcoma/primitive neuroectodermal tumor
    - Malignant pigmented neuroectodermal tumor of infancy (retinal anlage tumor)
- Paraganglionic tumors
  - Benign
    - Paraganglioma
  - Malignant
    - Malignant paraganglioma
- Extraskeletal osseous and cartilaginous tumors
  - Benign
    - Panniculitis ossificans and myositis ossificans
    - Fibroosseous pseudotumor of the digits
    - Fibrodysplasia ossificans progressiva
    - Extraskeletal chondroma or osteochondroma
    - Extraskeletal osteoma
  - Malignant
    - Extraskeletal chondrosarcoma
      - Well differentiated chondrosarcoma
      - Myxoid chondrosarcoma
      - Mesenchymal chondrosarcoma
    - Extraskeletal osteosarcoma
- Miscellaneous tumors
  - Benign
    - Congenital granular cell tumor
    - Tumoral calcinosis
    - Myxoma
      - Cutaneous
      - Intramuscular
    - Juxtaarticular myxoma
    - Aggressive angiomyxoma
    - Parachordoma
    - Amyloid tumor
    - Pleomorphic hyalinizing angiectatic tumor of soft parts
  - Intermediate
    - Ossifying fibromyxoid tumor of soft parts
    - Inflammatory myxohyaline tumor
  - Malignant
    - Synovial sarcoma
    - Alveolar soft part sarcoma
    - Epithelioid sarcoma
    - Desmoplastic small round cell tumor
    - Malignant extrarenal rhabdoid tumor

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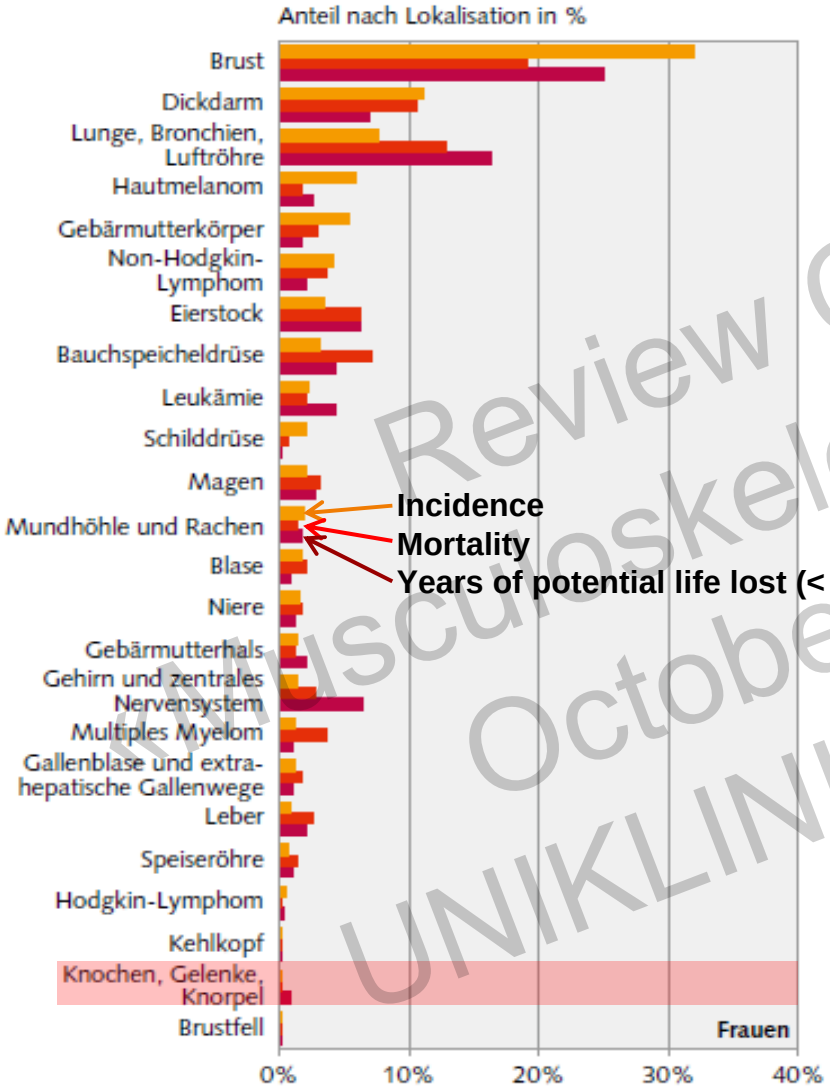
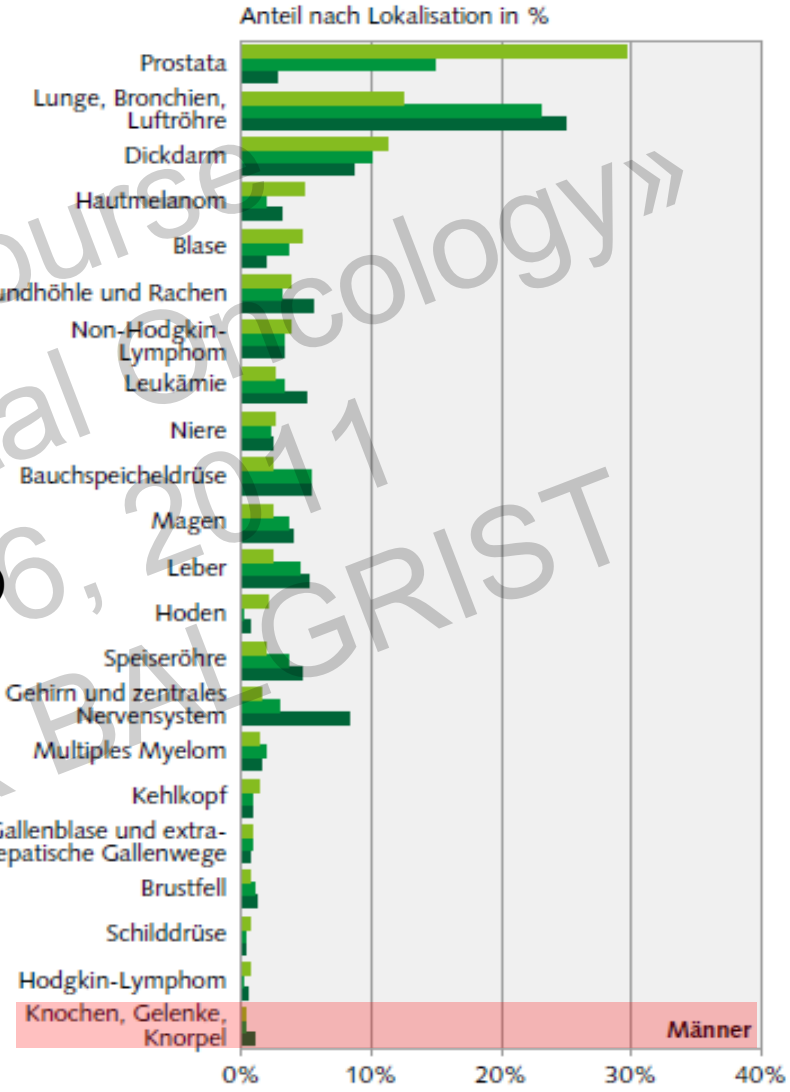
Malignancies in Switzerland



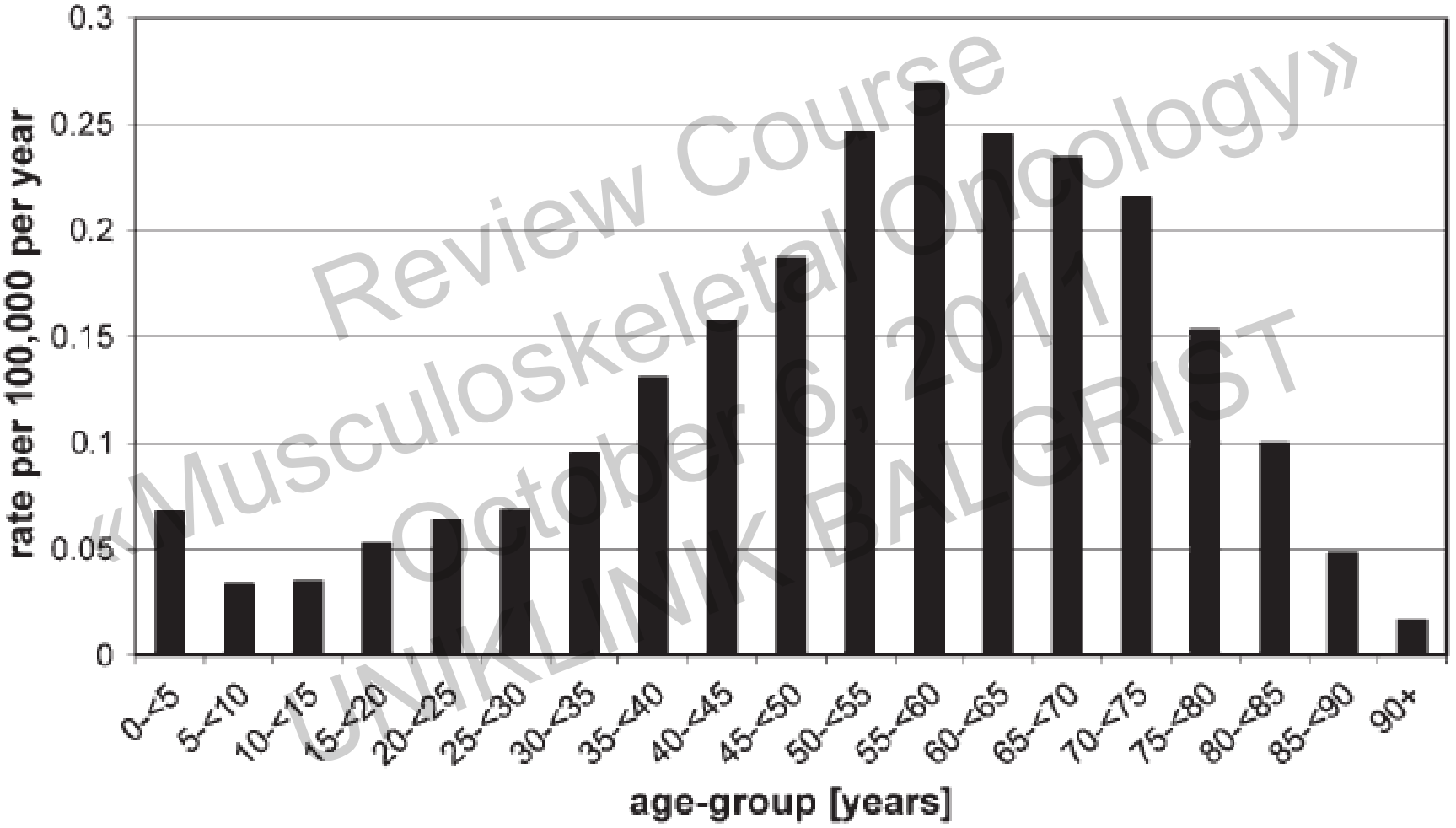


Bundesamt für Statistik. Krebs in der Schweiz, Neuchâtel 2011.

Soft Tissue Sarcomas (STS) in the USA

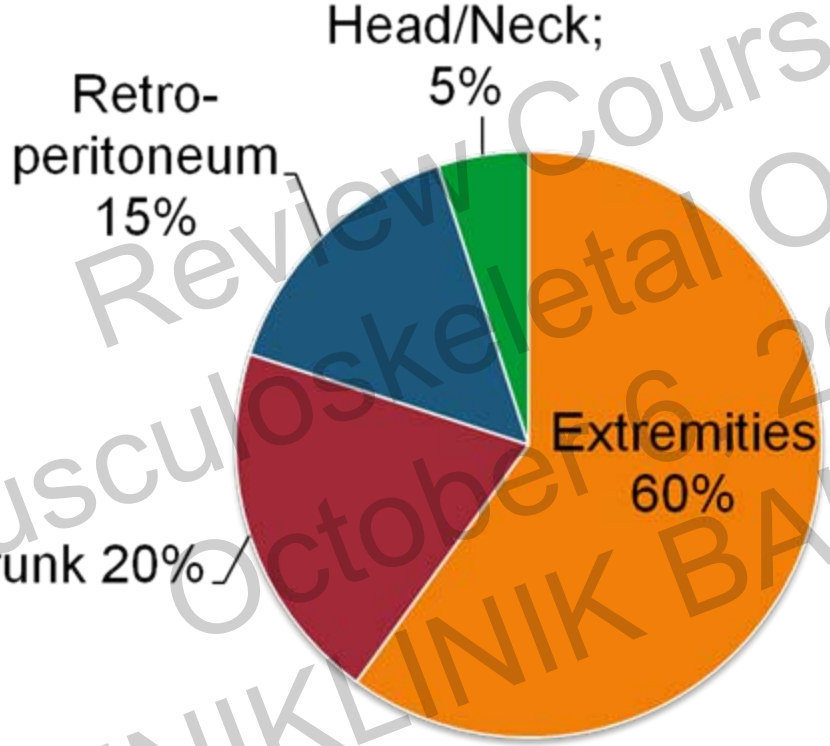
New cases	9'420 (2005)
Incidence [cases/100'000/y]	3.2
% of all cancers	<1%
Male : Female	1.4 : 1
% of all cancer deaths	0.61%
Benign lesions : STS	100:1

Age Distribution of STS



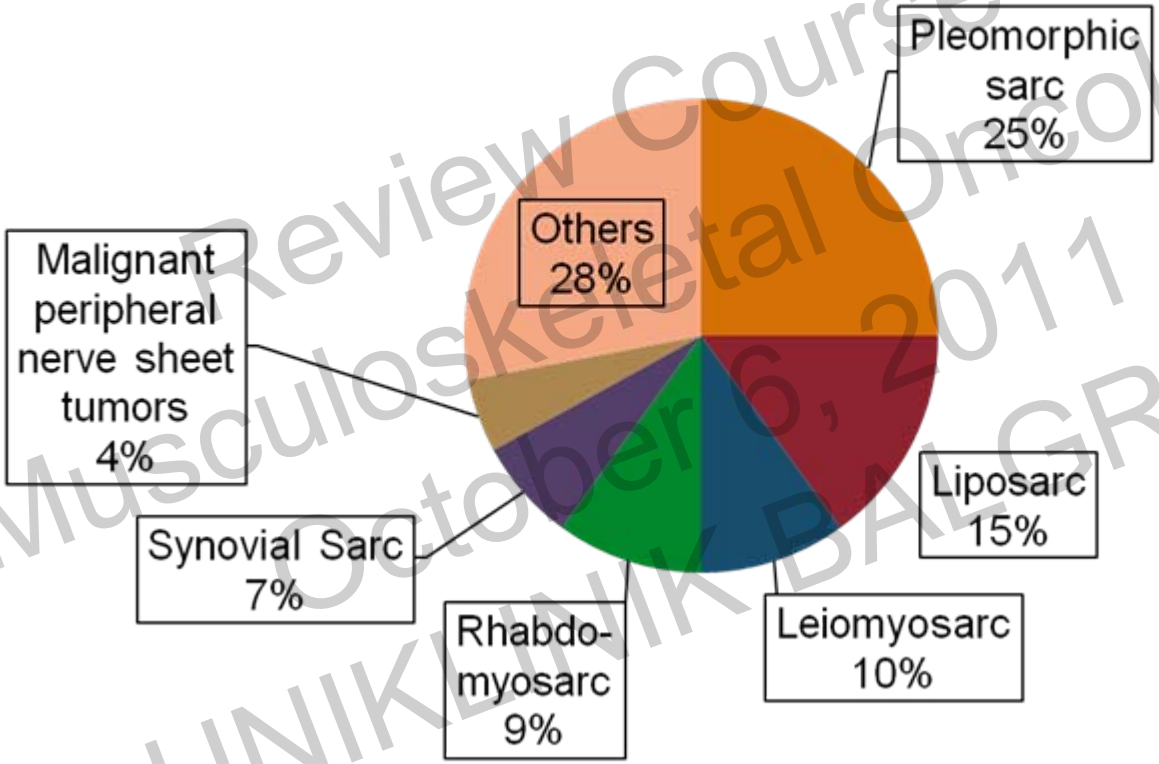
Wibmer C et al. Ann Oncol 2010;21:1106-1111.

Localisations STS



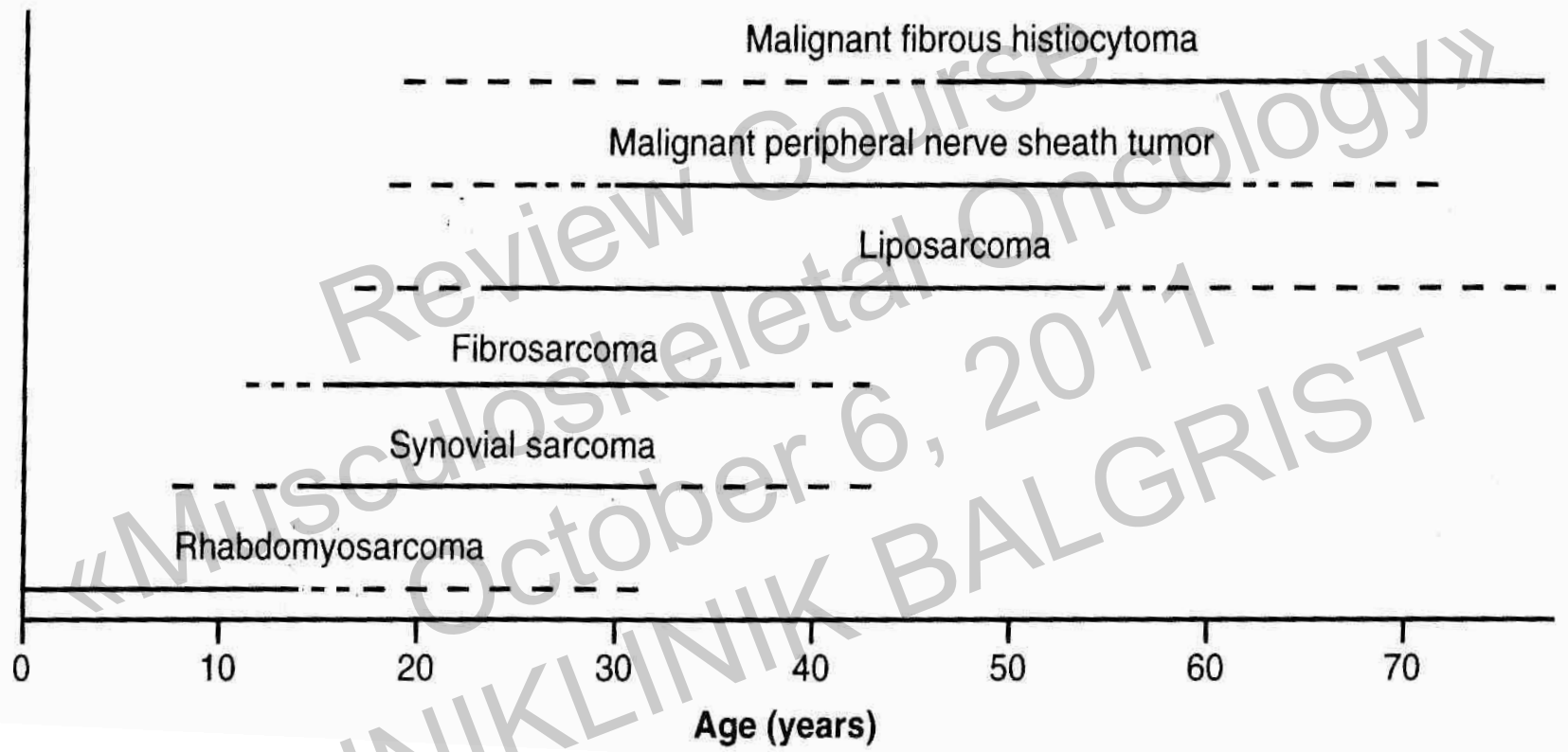
Quinn RH. Chapter 29. In: OKU Musculoskeletal Tumors 2. Schwarz HS Ed. 2007.AAOS, Rosemont/Il/USA.

Subtypes of STS



Quinn RH. Chapter 29. In: OKU Musculoskeletal Tumors 2. Schwarz HS Ed. 2007.AAOS, Rosemont/Il/USA.

Subtypes of STS and Age

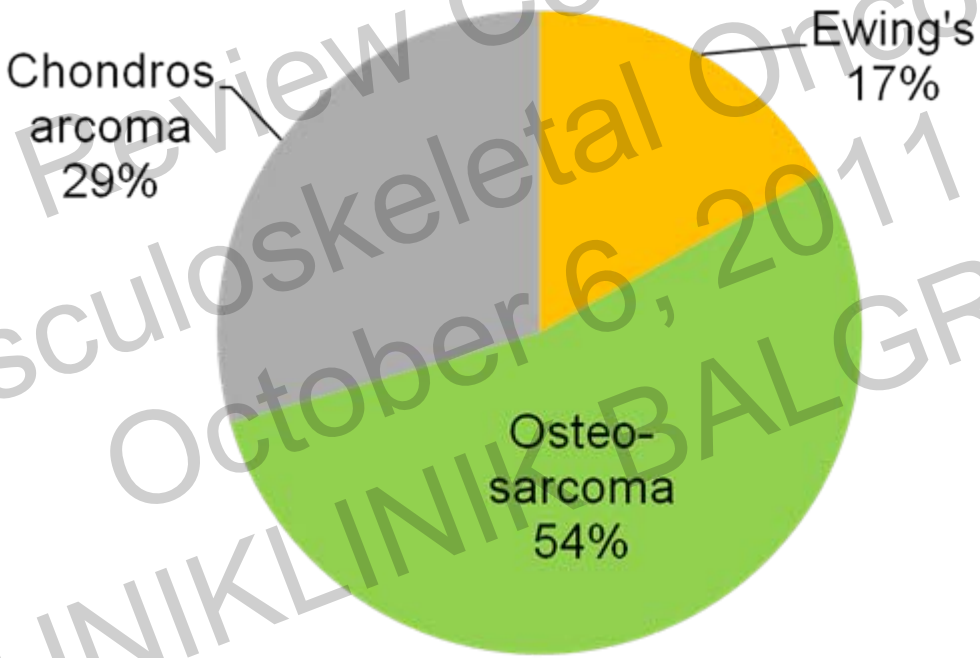


Weiss & Goldblum. In: Enzinger and Weiss's Soft Tissue Tumors. 4th ed. 2001. Mosby, St. Louis/USA. ISBN 0-232-01200-0.

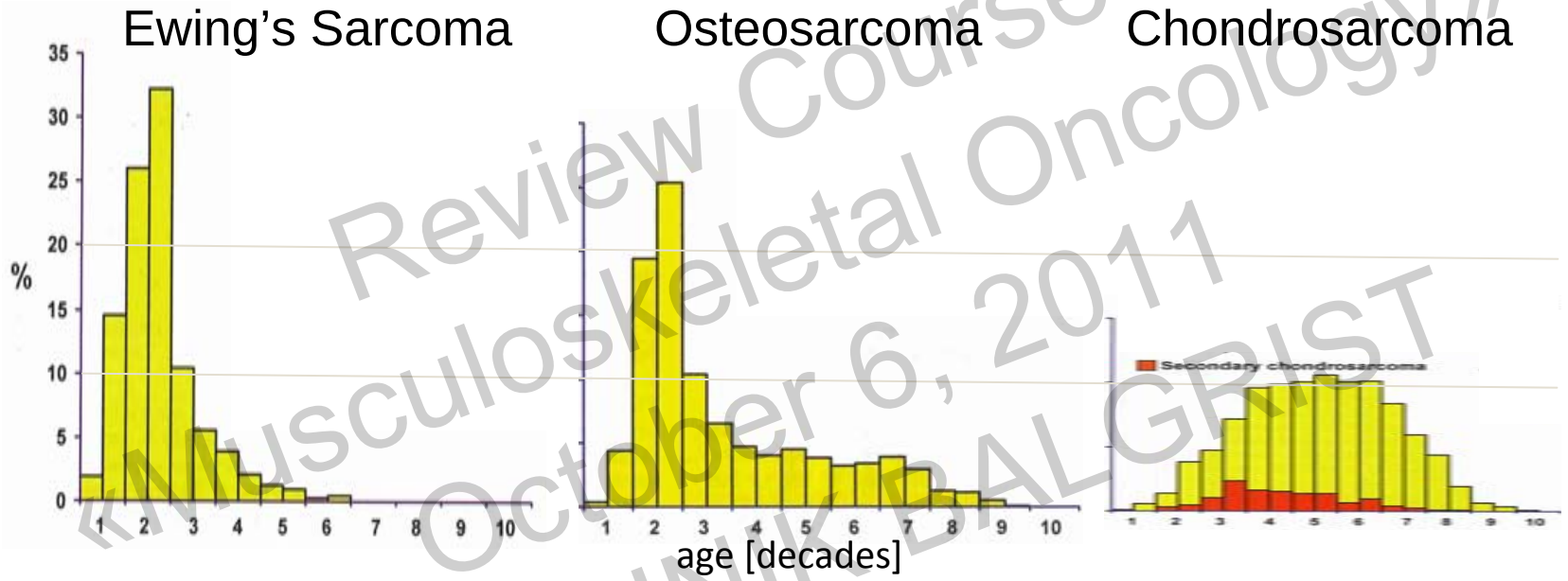
Sarcoma of Bone in the USA

New cases	2’650 (2010)
% of all cancers	<0.2%
Male : Female	1.4 : 1
Incidence	~10 cases / million /y
Incidence Osteosarcoma	4-5 cases / million /y

Subtypes of Primary Bone Sarcoma



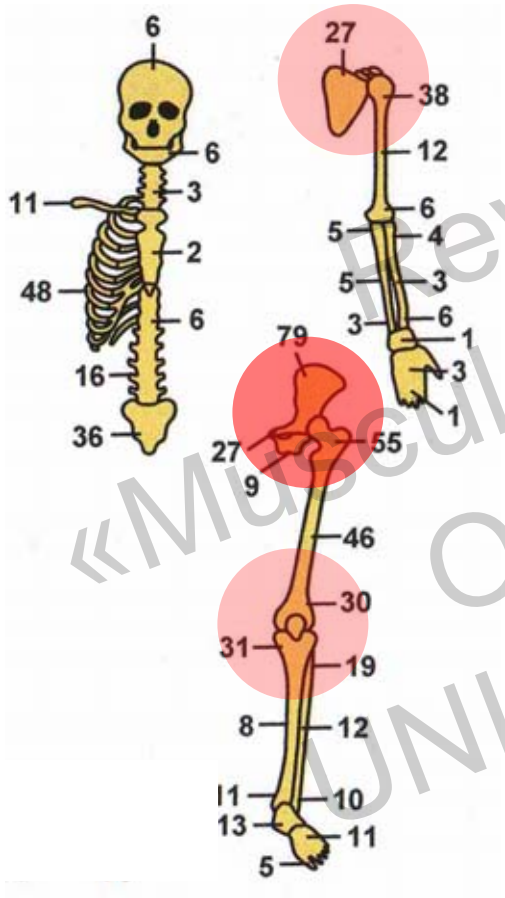
Age Distribution & Incidence of Primary Bone Sarcomas



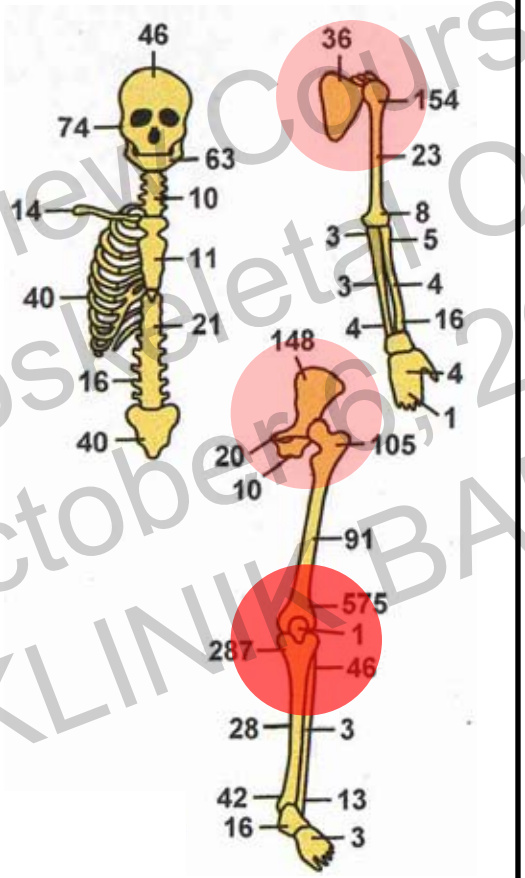
Unni KK & Inwards CY. 2010. In: Dahlins's Bone Tumors: General Aspects and Data of 10'165 cases. 6th ed. ISBN-13: 978-0-7817-6242-7.

Localisations of Primary Bone Sarcomas

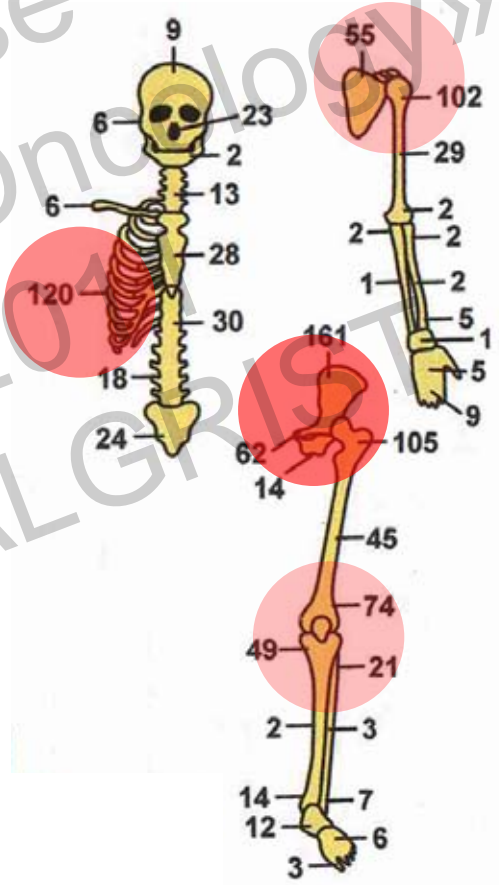
Ewing's Sarcoma



Osteosarcoma



Chondrosarcoma



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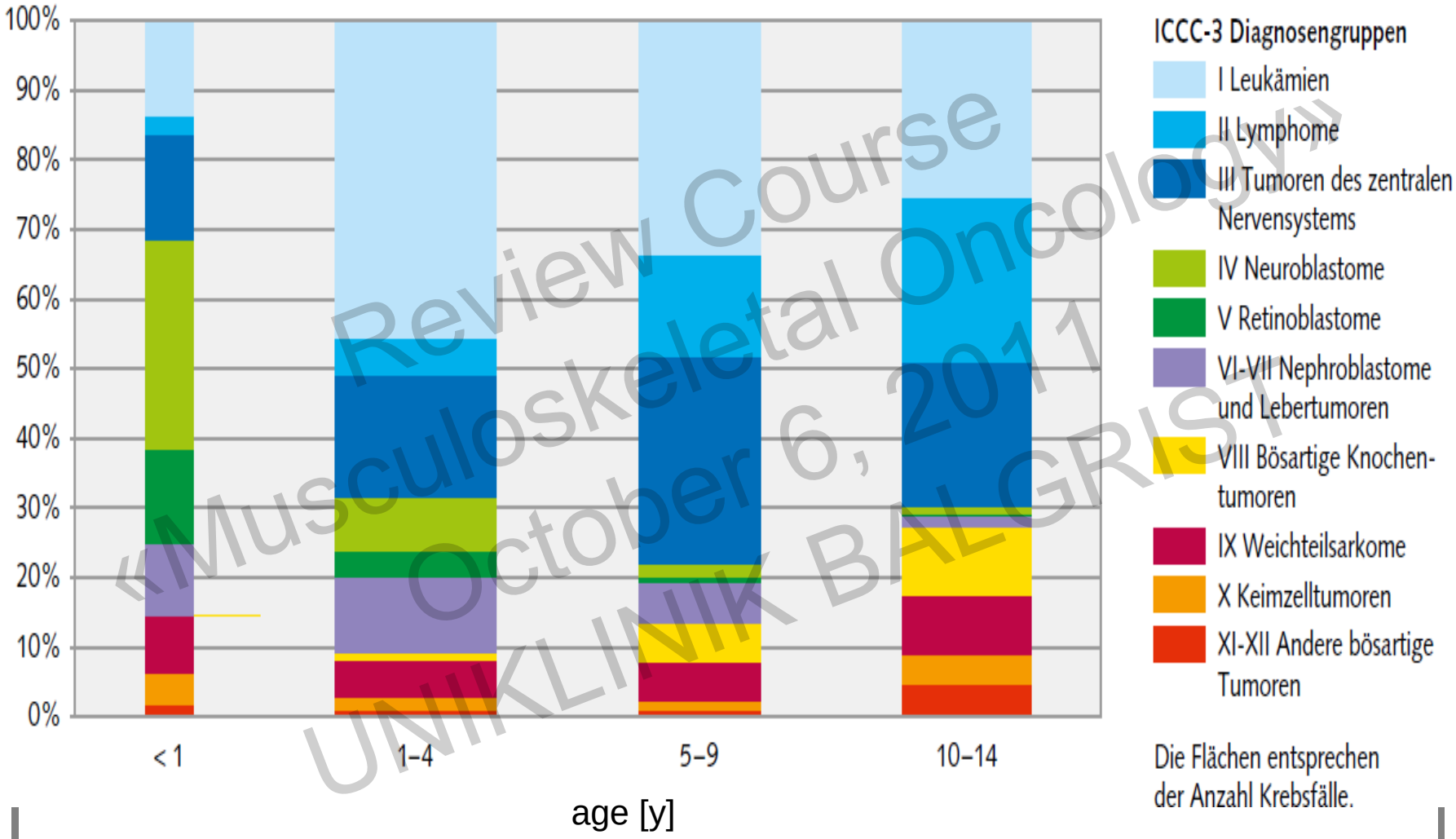
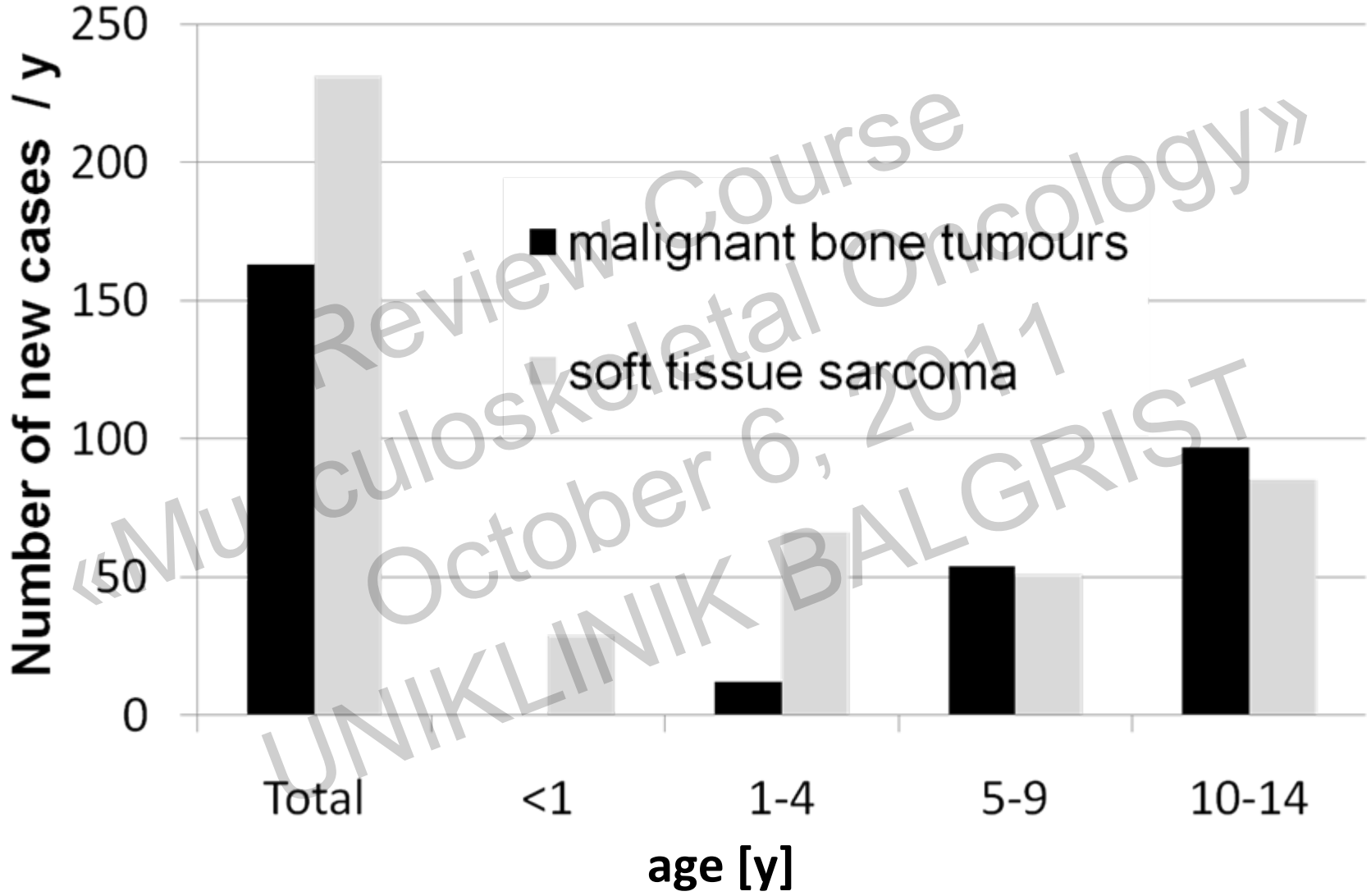
Pediatric Malignancies



Bundesamt für Statistik. Krebs in der Schweiz, Neuchâtel 2011.

Incidence of Pediatric Sarcoma in Switzerland



Bundesamt für Statistik. Krebs in der Schweiz, Neuchâtel 2011.

Cases treated in Switzeland 2005

	Total cases	Hospitals n	% treated in tertiary centers
Bone tumors	658	82	53
Soft tissue sarcoma	737	127	32

Medienmitteilung #0350-0711-90. EDI. Bundesamt für Statistik BFS. 2007.

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