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Introduction to sarcoma - epidemiology and etiology

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PGMO Sarcoma 12/12/2020

Sarcoma. Definition

Sarcoma is a heterogeneous group of rare malignant neoplasms arising from mesenchymal cells



- Altogether rare (<8 / 100,000 / year)
- <1% of all malignancies in adults, 15% of pediatric cancers

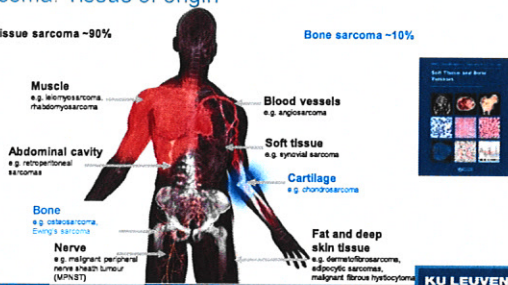
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Xiao H. Cancer Sarcoma Research 2013

Sarcoma. Tissue of origin

Soft tissue sarcoma ~90%

Bone sarcoma ~10%

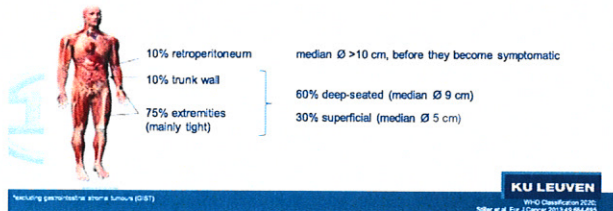


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Burningham et al. Clin Sarcoma Res. 2012;2(1):114
WHO Classification 2020

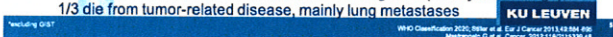
Epidemiology. Soft tissue sarcoma* (1)

- Incidence 4-5 cases / 100,000 / year
- Slight male predominance
- Anatomical site distribution



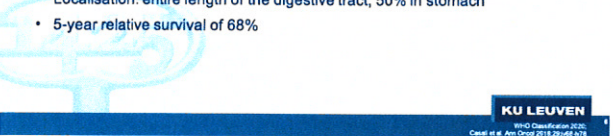
Epidemiology. Soft tissue sarcoma* (2)

- 110+ subtypes, most common:
 - undifferentiated pleomorphic sarcoma (UPS)
 - liposarcoma (LPS)
 - leiomyosarcoma (LMS)
 - myxofibrosarcoma (MFS)
 - synovial sarcoma (SynSa)
 - malignant peripheral nerve sheath tumours (MPNST)
- Median age at diagnosis 65 years, with variable age-related incidence:
 - exclusively in children: embryonal rhabdomyosarcoma
 - mainly in young adults: SynSa
 - elderly: UPS, LPS, LMS, MFS
- 5-year relative survival 58%
- ~10% of patients present with metastasis at diagnosis (mainly in the lungs).
1/3 die from tumor-related disease, mainly lung metastases



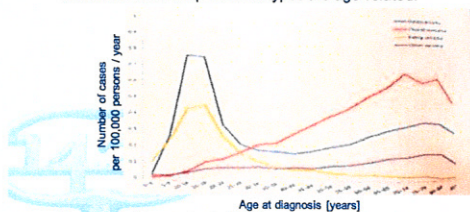
Epidemiology. Gastrointestinal stromal tumour (GIST)

- Incidence 1 / 100,000 / year (clinically relevant GIST)
 - much higher when including microGIST (< 1cm)
- Slight prevalence in males
- Median age 60-65 years
 - paediatric GIST represents a distinct subset
- Localisation: entire length of the digestive tract, 50% in stomach
- 5-year relative survival of 68%



Epidemiology. Bone sarcoma (1)

- Incidence: 0.75 case / 100,000 / year
- Incidence rates of specific subtypes are age-related:



- 5-year relative survival of 62%

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WHO Classification 2020
Siller et al. Eur J Cancer 2015; 55:484-492

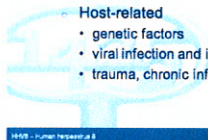
Epidemiology. Bone sarcoma (2)

	Osteosarcoma	Chondrosarcoma	Ewing's sarcoma
Incidence (per 100,000/year)	overall 0.3 0.8 - 1.1 at age 15-19y	0.2	0.3
Median age (years)	(bimodal distribution)	30 - 60	15
Male to female ratio	1.4:1	~1:1	1.5:1
Primary sites	extremity (younger patient) proportion of axial tumor increases with age	pelvis, proximal femur, ribs, humerus, tibia, and scapula	extremity bones (50%), pelvis, ribs, vertebra
Remarks	frequently associated with germline syndromes		

WHO Classification 2020

Etiology of sarcoma

- Sporadic >> genetic
- Cause mostly unknown
- Risk factors:
 - Environmental
 - radiation exposure (e.g. prior radiation therapy)
 - chemical carcinogens (e.g. dioxins in herbicides)
 - Host-related
 - genetic factors
 - viral infection and immunodeficiency (HHV8, EBV infection)
 - trauma, chronic inflammation



1505 - Tumor Sarcoma 18
1507 - Tumor Sarcoma

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WHO Classification 2020
Hu J. Surg Clin North Am. 2015;95(2):141-14

Etiology of sarcoma. Genetic factors

Genetic syndromes and their associated sarcomas		
Genetic syndrome	Mutation	Associated Sarcoma
L-Fraumeni syndrome	TP53	<ul style="list-style-type: none"> Osteosarcoma Rhabdomyosarcoma Liposarcoma
Retinoblastoma	RBI	<ul style="list-style-type: none"> Osteosarcoma Chondrosarcoma Festing sarcoma Uterine leiomyosarcoma Fibrosarcoma Rhabdomyosarcoma
Neurofibromatosis type 1	NF-1	<ul style="list-style-type: none"> Rhabdomyosarcoma Gastrointestinal stromal tumor Malignant peripheral nerve sheath tumor
Familial adenomatous polyposis	APC	Dermoid tumor
Familial gastrointestinal stromal tumors	KIT	Gastrointestinal stromal tumor
Crohn's syndrome	PCSK9A	
Crohn's Strahls syndrome	SDH	Gastrointestinal stromal tumor
Bloom's syndrome	BLM	Osteosarcoma
Weiner's syndrome	WRN	Osteosarcoma
Rothmund-Thomson syndrome	MCCO4	Osteosarcoma

Rothmund-Thomson syndrome	REC4D	Osteosarcoma
<p>TP53 tumor prone - R31, R91, retinoblastoma 1, M⁺, rasul bromista + age 1, APC adenomatous polyposis coli, R90A, R90B, R90C, R90D, R90E, R90F, R90G, R90H, R90I, R90J, R90K, R90L, R90M, R90N, R90O, R90P, R90Q, R90R, R90S, R90T, R90U, R90V, R90W, R90X, R90Y, R90Z, R90AA, R90AB, R90AC, R90AD, R90AE, R90AF, R90AG, R90AH, R90AI, R90AJ, R90AK, R90AL, R90AM, R90AN, R90AO, R90AP, R90AQ, R90AR, R90AS, R90AT, R90AU, R90AV, R90AW, R90AX, R90AY, R90AZ, R90BA, R90BB, R90BC, R90BD, R90BE, R90BF, R90BG, R90BH, R90BI, R90BJ, R90BK, R90BL, R90BM, R90BN, R90BO, R90BP, R90BQ, R90BR, R90BS, R90BT, R90BU, R90BV, R90BW, R90BX, R90BY, R90BZ, R90CA, R90CB, R90CC, R90CD, R90CE, R90CF, R90CG, R90CH, R90CI, R90CJ, R90CK, R90CL, R90CM, R90CN, R90CO, R90CP, R90CQ, R90CR, R90CS, R90CT, R90CU, R90CV, R90CW, R90CX, R90CY, R90CZ, R90DA, R90DB, R90DC, R90DD, R90DE, R90DF, R90DG, R90DH, R90DI, R90DJ, R90DK, R90DL, R90DM, R90DN, R90DO, R90DP, R90DQ, R90DR, R90DS, R90DT, R90DU, R90DV, R90DW, R90DX, R90DY, R90DZ, R90EA, R90EB, R90EC, R90ED, R90EE, R90EF, R90EG, R90EH, R90EI, R90EJ, R90EK, R90EL, R90EM, R90EN, R90EO, R90EP, R90EQ, R90ER, R90ES, R90ET, R90EU, R90EV, R90EW, R90EX, R90EY, R90EZ, R90FA, R90FB, R90FC, R90FD, R90FE, R90FF, R90FG, R90FH, R90FI, R90FJ, R90FK, R90FL, R90FM, R90FN, R90FO, R90FP, R90FQ, R90FR, R90FS, R90FT, R90FU, R90FV, R90FW, R90FX, R90FY, R90FZ, R90GA, R90GB, R90GC, R90GD, R90GE, R90GF, R90GG, R90GH, R90GI, R90GJ, R90GK, R90GL, R90GM, R90GN, R90GO, R90GP, R90GQ, R90GR, R90GS, R90GT, R90GU, R90GV, R90GW, R90GX, R90GY, R90GZ, R90HA, R90HB, R90HC, R90HD, R90HE, R90HF, R90HG, R90HH, R90HI, R90HJ, R90HK, R90HL, R90HM, R90HN, R90HO, R90HP, R90HQ, R90HR, R90HS, R90HT, R90HU, R90HV, R90HW, R90HX, R90HY, R90HZ, R90IA, R90IB, R90IC, R90ID, R90IE, R90IF, R90IG, R90IH, R90II, R90IJ, R90IK, R90IL, R90IM, R90IN, R90IO, R90IP, R90IQ, R90IR, R90IS, R90IT, R90IU, R90IV, R90IW, R90IX, R90IY, R90IZ, R90JA, R90JB, R90JC, R90JD, R90JE, R90JF, R90JG, R90JH, R90JI, R90JJ, R90JK, R90JL, R90JM, R90JN, 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R90ZZ</p>		



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