

**Classification of Soft Tissue and Bone Tumours based on ICD-O-3.2 (to be used for new registrations from 2020)**

WHO classification of tumours of soft tissue and bone: ICD-O coding	Classification ICD-O-3.2	COMMENTS	Topography
<b>WHO classification of soft tissue tumours</b>			
<b>Adipocytic tumours</b>			
<i>Intermediate (locally aggressive)</i>			
Atypical lipomatous tumour	8850/1	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
<i>Malignant</i>			
Well-differentiated liposarcoma, NOS	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Lipoma-like liposarcoma	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Inflammatory liposarcoma	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Sclerosing liposarcoma	8851/3	If you have the information about "MDM2+", note it in comment field	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
Dedifferentiated liposarcoma	8858/3	If you have the information about "MDM2+", note it in comment field	
Myxoid liposarcoma	8852/3	If you have the information about "DDIT3+", note it in comment field	
Pleomorphic liposarcoma	8854/3	If you have the information about "MDM2-" and "DDIT3-", note it in comment field	Extremities
Epithelioid liposarcoma	8854/3	If you have the information about "MDM2-" and "DDIT3-", note it in comment field	Extremities
Myxoid pleomorphic liposarcoma	8850/3	"8859/3" in comment, code does not yet exist in ICD-O-3.2, if you have the information about "TP53", note it in comment field	
Liposarcoma, NOS	8850/3	Needs to be justified in comment	It's important to have the initial topo to distinguish between 8851/3 and 8850/1
<b>Fibroblastic and myofibroblastic tumours</b>			
<i>Intermediate (locally aggressive)</i>			
Solitary fibrous tumour, benign	8815/0		
Palmar fibromatosis and plantar fibromatosis	8813/1		
Desmoid-type fibromatosis	8821/1		
Extra-abdominal desmoid	8821/1		
Abdominal fibromatosis	8822/1		
Lipofibromatosis	8851/1		
Giant cell fibroblastoma	8834/1		
<i>Intermediate (rarely metastasizing)</i>			
Dermatofibrosarcoma protuberans	8832/1		(C44._) exceptionnally C49
Pigmented dermatofibrosarcoma protuberans (Bednar tumour)	8833/1		(C44._) exceptionnally C49
Dermatofibrosarcoma protuberans, fibrosarcomatous	8832/3		(C44._) exceptionnally C49
Solitary fibrous tumour, NOS	8815/1		
Inflammatory myofibroblastic tumour	8825/1		
(Low-grade) myofibroblastic sarcoma	8825/3		
Superficial CD34-positive fibroblastic tumour	8810/1	"Superficial CD34-positive fibroblastic tumour" in comment, new in WHO 2020 (futur ICD-O-4)	
Myxoinflammatory fibroblastic sarcoma	8811/1	"Myxoinflammatory fibroblastic sarcoma" in comment	
Infantile fibrosarcoma	8814/3		
<i>Malignant</i>			
Solitary fibrous tumour, malignant	8815/3		
Fibrosarcoma, NOS	8810/3		
Myxofibrosarcoma	8811/3		
Low-grade fibromyxoid sarcoma	8840/3		
Sclerosing epithelioid fibrosarcoma	8840/3		

**So-called fibrohistiocytic tumours***Intermediate (rarely metastasizing)*

Plexiform fibrohistiocytic tumour	8835/1
Giant cell tumour of soft parts	9251/1

*Malignant*

Malignant tenosynovial giant cell tumour	9252/3
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(C49..)

**Vascular tumours***Intermediate (locally aggressive)*

Kaposiform haemangioendothelioma	9130/1
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*Intermediate (rarely metastasizing)*

Retiform haemangioendothelioma	9136/1
Composite haemangioendothelioma	9136/1
Neuroendocrine composite haemangioendothelioma	9136/1
Papillary intralymphatic angioendothelioma	9135/1
Pseudomyogenic (epithelioid sarcoma-like) haemangioendothelioma	9138/1
Kaposi sarcoma	9140/3
Classic indolent Kaposi sarcoma	9140/3
Endemic African Kaposi sarcoma	9140/3
AIDS-associated Kaposi sarcoma	9140/3
Iatrogenic Kaposi sarcoma	9140/3

*Malignant*

Epithelioid haemangioendothelioma NOS	9133/3
Epithelioid haemangioendothelioma with WWTR1-CAMTA1 fusion	9133/3
Epithelioid haemangioendothelioma with YAP1-TFE3 fusion	9133/3
Angiosarcoma	9120/3

**Pericytic (perivascular) tumours***Intermediate*

Glomangiomas	8711/1
Glomus tumour of uncertain malignant potential	8711/1
Myofibromatosis	8824/1
Infantile myofibromatosis	8824/1

*Malignant*

Glomus tumour, malignant	8711/3
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**Smooth muscle tumours***Intermediate*

EBV-associated smooth muscle tumour, smooth muscle tumour NOS	8897/1
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"EBV-associated smooth muscle tumour " in comment,  
new in WHO 2020 (futur ICD-O-4)*Malignant*

Leiomyosarcoma, NOS	8890/3
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**Skeletal muscle tumours***Malignant*

Embryonal rhabdomyosarcoma, NOS	8910/3
Embryonal rhabdomyosarcoma, pleomorphic	8910/3
Alveolar rhabdomyosarcoma	8920/3
Pleomorphic rhabdomyosarcoma, NOS	8901/3
Spindle cell rhabdomyosarcoma	8912/3
Congenital spindle cell rhabdomyosarcoma (with VGLL2/NCOA2/CITED2 rearrangements)	8912/3
MYOD1-mutant spindle cell / sclerosing rhabdomyosarcoma	8912/3
Intraosseous spindle cell rhabdomyosarcoma (with TFCP2/NCOA2 rearrangements)	8912/3
Ectomesenchymoma	8921/3
Rhabdomyosarcoma, NOS	8900/3

**Chondro-osseous tumours***Malignant*

Osteosarcoma, extraskeletal	9180/3
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**Peripheral nerve sheath tumours***Malignant*

Malignant peripheral nerve sheath tumour (MPNST), NOS	9540/3	
Malignant melanotic nerve sheath tumour	9540/3	"Malignant melanotic nerve sheath tumour " in comment, new in WHO 2020 (futur ICD-O-4)
Malignant peripheral nerve sheath tumour (MPNST) with skeletal muscle differentiation	9561/3	
Malignant Triton tumour	9561/3	
Malignant peripheral nerve sheath tumour (MPNST), epithelioid	9542/3	
Granular cell tumour, malignant	9580/3	
Perineurioma, malignant	9571/3	

**Tumours of uncertain differentiation***Intermediate (locally aggressive)*

Haemosiderotic fibrolipomatous tumour	8811/1	"Haemosiderotic fibrolipomatous tumour" in comment
Angiomyolipoma, epithelioid	8860/1	

*Intermediate (rarely metastasizing)*

Atypical fibroxanthoma	8830/1
Angiomatoid fibrous histiocytoma	8836/1
Ossifying fibromyxoid tumour, NOS	8842/0
Mixed tumour, NOS	8940/0
Mixed tumour, malignant	8940/3
Myoepithelioma, NOS	8982/0

*Malignant*

Phosphaturic mesenchymal tumour, malignant; Mesenchymoma, malignant	8990/3	
NTRK-rearranged spindle cell neoplasm (emerging)	8800/3	"NTRK-rearranged spindle cell neoplasm (emerging), malignant" in comment
Synovial sarcoma, NOS	9040/3	
Synovial sarcoma, poorly differentiated	9040/3	
Synovial sarcoma, spindle cell	9041/3	
Synovial sarcoma, biphasic	9043/3	

Epithelioid sarcoma	8804/3	
Proximal or large cell epithelioid sarcoma	8804/3	
Classic epithelioid sarcoma	8804/3	
Alveolar soft part sarcoma	9581/3	
Clear cell sarcoma, NOS	9044/3	
Clear cell sarcoma of kidney	8964/3	(C64._)
Extraskeletal myxoid chondrosarcoma	9231/3	
Desmoplastic small round cell tumour	8806/3	
Rhabdoid tumour, NOS	8963/3	Outside the central nervous system (renal or extrarenal)
Atypical teratoid/rhabdoid tumour	9508/3	(C71._)
Perivascular epithelioid tumor (PEComa), malignant	8714/3	
Intimal sarcoma	9137/3	
Ossifying fibromyxoid tumour, malignant	8842/3	
Myoepithelial carcinoma	8982/3	

#### Gastrointestinal stromal tumours

Gastrointestinal stromal tumour	8936/3	Mitotic rate should be added in comment
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#### Endometrial stromal sarcoma

##### Malignant

Endometrial stromal sarcoma, high grade, NOS	8930/3	(C54.1)
Endometrial stromal sarcoma, low grade	8931/3	(C54.1)

#### Unclassified and poorly characterised tumours

##### Malignant

Stromal sarcoma, NOS	8935/3	Needs to be justified in comment
Stromal tumour of uncertain malignant potential, NOS	8935/1	Needs to be justified in comment
Sarcoma, NOS	8800/3	Needs to be justified in comment

#### WHO classification of undifferentiated small round cell sarcomas of bone and soft tissue

Ewing sarcoma	9364/3	
Round cell sarcoma with EWSR1-non-ETS fusions	8803/3	9366/3 in comment. Code does not yet exist in ICD-O-3.2
CIC-rearranged sarcoma	8803/3	9367/3 in comment. Code does not yet exist in ICD-O-3.2
Sarcoma with BCOR genetic alterations	8803/3	9368/3 in comment. Code does not yet exist in ICD-O-3.2

**WHO classification of bone tumours****Chondrogenic tumours***Intermediate (locally aggressive)*

Synovial chondromatosis 9220/1

Central atypical cartilaginous tumour 9222/1

Secondary peripheral atypical cartilaginous tumour 9222/1

*Malignant*

Central chondrosarcoma, grade 1 9222/3

Secondary chondrosarcoma, grade 1 9222/3

Central chondrosarcoma, grade 2 9220/3

Secondary peripheral chondrosarcoma, grade 2 9220/3

Central chondrosarcoma, grade 3 9220/3

Secondary peripheral chondrosarcoma, grade 3 9220/3

Periosteal chondrosarcoma 9221/3

Clear cell chondrosarcoma 9242/3

Mesenchymal chondrosarcoma 9240/3

Dedifferentiated chondrosarcoma 9243/3

(C40.\_, C41.\_) exceptionally soft tissue

(C40.\_, C41.\_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 &amp; 9222/1 (C40.\_, C41.\_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 &amp; 9222/1

(C40.\_, C41.\_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 &amp; 9222/1

(C40.\_, C41.\_) exceptionally soft tissue, use the initial topo to distinguish between 9222/3 &amp; 9222/1 (C40.\_, C41.\_) exceptionally soft tissue

(C40.\_, C41.\_) also possible in soft tissue, and intracranial sites

(C40.\_, C41.\_) exceptionally soft tissue

**Osteogenic tumours***Intermediate (locally aggressive)*

Osteoblastoma, NOS 9200/1

*Malignant*

Low-grade central osteosarcoma 9187/3

Osteosarcoma, NOS 9180/3

Conventional osteosarcoma 9180/3

Telangiectatic osteosarcoma 9183/3

Small cell osteosarcoma 9185/3

Parosteal osteosarcoma 9192/3

Periosteal osteosarcoma 9193/3

High grade surface osteosarcoma 9194/3

Secondary osteosarcoma 9184/3

(C40.\_, C41.\_) exceptionally soft tissue

**Fibroblastic tumours***Intermediate (locally aggressive)*

Desmoplastic fibroma 8823/1

*Malignant*

Fibrosarcoma, NOS 8810/3

**Vascular tumours***Intermediate (locally aggressive)*

Epithelioid haemangioma 9125/0

*Malignant*

Epithelioid haemangioendothelioma NOS 9133/3

Angiosarcoma 9120/3

**Osteoclastic giant cell-rich tumours***Intermediate (locally aggressive, rarely metastasizing)*

Giant cell tumour of bone 9250/1

*Malignant*

Giant cell tumour of bone, malignant 9250/3

**Notochordal tumours***Malignant*

Conventional chordoma	9370/3
Poorly differentiated chordoma	9370/3
Chondroid chordoma	9370/3
Dedifferentiated chordoma	9372/3

**Other mesenchymal bone tumours***Intermediate (locally aggressive)*

Osteofibrous dysplasia-like adamantinoma	9261/1
Fibrocartilaginous mesenchymoma	8990/1

*Malignant*

Adamantinoma of long bones	9261/3	(C40._)
Dedifferentiated adamantinoma	9261/3	(C40._)
Leiomyosarcoma NOS	8890/3	(C40._, C41._)
Pleomorphic sarcoma, undifferentiated	8802/3	(C40._, C41._)

**WHO classification of bone tumours and soft tissue tumours of uncertain differentiation****Tumours of uncertain differentiation***Malignant*

Epithelioid sarcoma, NOS, undifferentiated	8804/3	After exclusion	
Undifferentiated sarcoma	8805/3	After exclusion	
Spindle cell sarcoma, undifferentiated	8801/3	After exclusion	
Pleomorphic cell sarcoma, undifferentiated	8802/3	After exclusion	(C44._) for the pleomorphic dermal sarcoma
Round cell sarcoma, undifferentiated	8803/3	After exclusion	
Sarcoma, NOS	8800/3	Needs to be justified in comment	

*In grey: codes and / or names of entities which should be avoided in favour of a more specific code*