Soft Tissue and Bone Tumours/ Foreword
/ WHO classification of tumours of soft tissue and bone: ICD-O coding

Headings

WHO classification of soft tissue tumours

Adipocytic tumours

Benign

8850/0 Lipoma NOS

- 8856/0 Intramuscular lipoma
- Chondrolipoma

Lipomatosis

- Diffuse lipomatosis
- Multiple symmetrical lipomatosis
- Pelvic lipomatosis
- Steroid lipomatosis
- HIV lipodystrophy

Lipomatosis of nerve

8881/0 Lipoblastomatosis

- Localized (lipoblastoma)
- Diffuse (lipoblastomatosis)

8861/0 Angiolipoma NOS

- Cellular angiolipoma

8890/0 Myolipoma

8862/0 Chondroid lipoma

8857/0 Spindle cell lipoma

8857/0 Atypical spindle cell / pleomorphic lipomatous tumour

8880/0 Hibernoma

Intermediate (locally aggressive)

8850/1 Atypical lipomatous tumour

Malignant

8851/3 Liposarcoma, well-differentiated, NOS

8851/3 Lipoma-like liposarcoma

8851/3 Inflammatory liposarcoma

8851/3 Sclerosing liposarcoma

8858/3 Dedifferentiated liposarcoma

8852/3 Myxoid liposarcoma

8854/3 Pleomorphic liposarcoma

Epithelioid liposarcoma

8859/3* Myxoid pleomorphic liposarcoma

Fibroblastic and myofibroblastic tumours

Benign

8828/0 Nodular fasciitis

Intravascular fasciitis

Cranial fasciitis

8828/0 Proliferative fasciitis

8828/0 Proliferative myositis

Myositis ossificans and fibro-osseous pseudotumour of digits

Ischaemic fasciitis

8820/0 Elastofibroma

8992/0 Fibrous hamartoma of infancy

Fibromatosis colli

Juvenile hyaline fibromatosis

Inclusion body fibromatosis

8813/0 Fibroma of tendon sheath

8810/0 Desmoplastic fibroblastoma

8825/0 Myofibroblastoma

8816/0 Calcifying aponeurotic fibroma

EWSR1-SMAD3–positive fibroblastic tumour (emerging)

8826/0 Angiomyofibroblastoma

9160/0 Cellular angiofibroma

9160/0 Angiofibroma

8810/0 Nuchal fibroma

8811/0 Acral fibromyxoma

8810/0 Gardner fibroma

Intermediate (locally aggressive)

8815/0 Solitary fibrous tumour, benign

8813/1 Palmar/plantar-type fibromatosis

8821/1 Desmoid-type fibromatosis

8821/1 Extra-abdominal desmoid

8822/1 Abdominal fibromatosis

8851/1 Lipofibromatosis

8834/1 Giant cell fibroblastoma

Intermediate (rarely metastasizing)

8832/1 Dermatofibrosarcoma protuberans NOS

8833/1 Pigmented dermatofibrosarcoma protuberans

8832/3 Dermatofibrosarcoma protuberans, fibrosarcomatous

Myxoid dermatofibrosarcoma protuberans

Dermatofibrosarcoma protuberans with myoid differentiation

Plaque-like dermatofibrosarcoma protuberans

8815/1 Solitary fibrous tumour NOS

Fat-forming (lipomatous) solitary fibrous tumour

Giant cell–rich solitary fibrous tumour

8825/1 Inflammatory myofibroblastic tumour

Epithelioid inflammatory myofibroblastic sarcoma

8825/3 Myofibroblastic sarcoma

8810/1 Superficial CD34-positive fibroblastic tumour

8811/1 Myxoinflammatory fibroblastic sarcoma

8814/3 Infantile fibrosarcoma

Malignant

8815/3 Solitary fibrous tumour, malignant

8810/3 Fibrosarcoma NOS

8811/3 Myxofibrosarcoma

Epithelioid myxofibrosarcoma

8840/3 Low-grade fibromyxoid sarcoma

8840/3 Sclerosing epithelioid fibrosarcoma

So-called fibrohistiocytic tumours

Benign

9252/0 Tenosynovial giant cell tumour NOS

9252/1 Tenosynovial giant cell tumour, diffuse

8831/0 Deep benign fibrous histiocytoma

Intermediate (rarely metastasizing)

8835/1 Plexiform fibrohistiocytic tumour

9251/1 Giant cell tumour of soft parts

Malignant

9252/3 Malignant tenosynovial giant cell tumour

Vascular tumours

Benign

9120/0 Haemangioma NOS

9132/0 Intramuscular haemangioma

9123/0 Arteriovenous haemangioma

9122/0 Venous haemangioma

9125/0 Epithelioid haemangioma

Cellular epithelioid haemangioma

Atypical epithelioid haemangioma

9170/0 Lymphangioma NOS

Lymphangiomatosis

9173/0 Cystic lymphangioma

9161/0 Acquired tufted haemangioma

Intermediate (locally aggressive)

9130/1 Kaposiform haemangioendothelioma

Intermediate (rarely metastasizing)

9136/1 Retiform haemangioendothelioma

9135/1 Papillary intralymphatic angioendothelioma

9136/1 Composite haemangioendothelioma

Neuroendocrine composite haemangioendothelioma

9140/3 Kaposi sarcoma

Classic indolent Kaposi sarcoma

Endemic African Kaposi sarcoma

AIDS-associated Kaposi sarcoma

Iatrogenic Kaposi sarcoma

9138/1 Pseudomyogenic (epithelioid sarcoma–like) haemangioendothelioma

Malignant

9133/3 Epithelioid haemangioendothelioma NOS

Epithelioid haemangioendothelioma with *WWTR1-CAMTA1* fusion

Epithelioid haemangioendothelioma with *YAP1-TFE3* fusion

9120/3 Angiosarcoma

Pericytic (perivascular) tumours

Benign and intermediate

8711/0 Glomus tumour NOS

8712/0 Glomangioma

8713/0 Glomangiomyoma

8711/1 Glomangiomatosis

8711/1 Glomus tumour of uncertain malignant potential

8824/0 Myopericytoma

8824/1 Myofibromatosis

8824/0 Myofibroma

8824/1 Infantile myofibromatosis

8894/0 Angioleiomyoma

Malignant

8711/3 Glomus tumour, malignant

Smooth muscle tumours

Benign and intermediate

8890/0 Leiomyoma NOS

8897/1 Smooth muscle tumour of uncertain malignant potential

Malignant

8890/3 Leiomyosarcoma NOS

Skeletal muscle tumours

Benign

8900/0 Rhabdomyoma NOS

- 8903/0 Fetal rhabdomyoma
- 8904/0 Adult rhabdomyoma
- 8905/0 Genital rhabdomyoma

Malignant

8910/3 Embryonal rhabdomyosarcoma NOS

- 8910/3 Embryonal rhabdomyosarcoma, pleomorphic

8920/3 Alveolar rhabdomyosarcoma

8901/3 Pleomorphic rhabdomyosarcoma NOS

8912/3 Spindle cell rhabdomyosarcoma

- Congenital spindle cell rhabdomyosarcoma with *VGLL2/NCOA2/CITED2* rearrangements
- *MYOD1*-mutant spindle cell / sclerosing rhabdomyosarcoma
- Intraosseous spindle cell rhabdomyosarcoma (with *TFCP2/NCOA2* rearrangements)

8921/3 Ectomesenchymoma

Gastrointestinal stromal tumours

8936/3 Gastrointestinal stromal tumour

Chondro-osseous tumours

Benign

9220/0 Chondroma NOS

Chondroblastoma-like soft tissue chondroma

Malignant

9180/3 Osteosarcoma, extraskeletal

Peripheral nerve sheath tumours

Benign

9560/0 Schwannoma NOS

9560/0 Ancient schwannoma

9560/0 Cellular schwannoma

9560/0 Plexiform schwannoma

Epithelioid schwannoma

Microcystic/reticular schwannoma

9540/0 Neurofibroma NOS

Ancient neurofibroma

Cellular neurofibroma

Atypical neurofibroma

9550/0 Plexiform neurofibroma

9571/0 Perineurioma NOS

Reticular perineurioma

Sclerosing perineurioma

9580/0 Granular cell tumour NOS

9562/0 Nerve sheath myxoma

9570/0 Solitary circumscribed neuroma

Plexiform solitary circumscribed neuroma

9530/0 Meningioma NOS

Benign triton tumour / neuromuscular choristoma

9563/0 Hybrid nerve sheath tumour

Perineurioma/schwannoma

Schwannoma/neurofibroma

Perineurioma/neurofibroma

Malignant

9540/3 Malignant peripheral nerve sheath tumour NOS

9542/3 Malignant peripheral nerve sheath tumour, epithelioid

9540/3 Malignant melanotic nerve sheath tumour

9580/3 Granular cell tumour, malignant

9571/3 Perineurioma, malignant

Tumours of uncertain differentiation

Benign

8840/0 Myxoma NOS

Cellular myxoma

8841/0 Aggressive angiomyxoma

8802/1 Pleomorphic hyalinizing angiectatic tumour

8990/0 Phosphaturic mesenchymal tumour NOS

8714/0 Perivascular epithelioid tumour, benign

8860/0 Angiomyolipoma

Intermediate (locally aggressive)

8811/1 Haemosiderotic fibrolipomatous tumour

8860/1 Angiomyolipoma, epithelioid

Intermediate (rarely metastasizing)

8830/1 Atypical fibroxanthoma

8836/1 Angiomatoid fibrous histiocytoma

8842/0 Ossifying fibromyxoid tumour NOS

8940/0 Mixed tumour NOS

8940/3 Mixed tumour, malignant, NOS

8982/0 Myoepithelioma NOS

Malignant

8990/3 Phosphaturic mesenchymal tumour, malignant

NTRK-rearranged spindle cell neoplasm (emerging)

9040/3 Synovial sarcoma NOS

9041/3 Synovial sarcoma, spindle cell

9043/3 Synovial sarcoma, biphasic

Synovial sarcoma, poorly differentiated

8804/3 Epithelioid sarcoma

Proximal or large cell epithelioid sarcoma

Classic epithelioid sarcoma

9581/3 Alveolar soft part sarcoma

9044/3 Clear cell sarcoma NOS

9231/3 Extraskeletal myxoid chondrosarcoma

8806/3 Desmoplastic small round cell tumour

8963/3 Rhabdoid tumour NOS

8714/3 Perivascular epithelioid tumour, malignant

9137/3 Intimal sarcoma

8842/3 Ossifying fibromyxoid tumour, malignant

8982/3 Myoepithelial carcinoma

8805/3 Undifferentiated sarcoma

8801/3 Spindle cell sarcoma, undifferentiated

8802/3 Pleomorphic sarcoma, undifferentiated

8803/3 Round cell sarcoma, undifferentiated

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

9364/3 Ewing sarcoma

9366/3* Round cell sarcoma with *EWSR1*–non-ETS fusions

9367/3* *CIC*-rearranged sarcoma

9368/3* Sarcoma with *BCOR* genetic alterations

WHO classification of bone tumours

Chondrogenic tumours

Benign

9213/0 Subungual exostosis

9212/0 Bizarre parosteal osteochondromatous proliferation

9221/0 Periosteal chondroma

9220/0 Enchondroma

9210/0 Osteochondroma

9230/0 Chondroblastoma NOS

9241/0 Chondromyxoid fibroma

9211/0 Osteochondromyxoma

Intermediate (locally aggressive)

9220/1 Synovial chondromatosis

9222/1 Atypical cartilaginous tumour

Malignant

9222/3* Chondrosarcoma, grade 1

9220/3 Chondrosarcoma, grade 2

9220/3 Chondrosarcoma, grade 3

9221/3 Periosteal chondrosarcoma

9242/3 Clear cell chondrosarcoma

9240/3 Mesenchymal chondrosarcoma

9243/3 Dedifferentiated chondrosarcoma

Osteogenic tumours

Benign

9180/0 Osteoma

9191/0 Osteoid osteoma

Intermediate (locally aggressive)

9200/1* Osteoblastoma NOS

Malignant

9187/3 Low-grade central osteosarcoma

9180/3 Osteosarcoma NOS

- Conventional osteosarcoma
- Telangiectatic osteosarcoma
- Small cell osteosarcoma

9192/3 Parosteal osteosarcoma

9193/3 Periosteal osteosarcoma

9194/3 High-grade surface osteosarcoma

9184/3 Secondary osteosarcoma

Fibrogenic tumours

Intermediate (locally aggressive)

8823/1 Desmoplastic fibroma

Malignant

8810/3 Fibrosarcoma NOS

Vascular tumours of bone

Benign

9120/0 Haemangioma NOS

Intermediate (locally aggressive)

9125/0 Epithelioid haemangioma

Malignant

9133/3 Epithelioid haemangioendothelioma NOS

9120/3 Angiosarcoma

Osteoclastic giant cell–rich tumours

Benign

9260/0 Aneurysmal bone cyst

8830/0 Non-ossifying fibroma

Intermediate (locally aggressive, rarely metastasizing)

9250/1 Giant cell tumour of bone

Malignant

9250/3 Giant cell tumour of bone, malignant

Notochordal tumours

Benign

9370/0 Benign notochordal tumour

Malignant

9370/3 Conventional chordoma

Chondroid chordoma

9370/3 Poorly differentiated chordoma

9372/3 Dedifferentiated chordoma

Other mesenchymal tumours of bone

Benign

Chondromesenchymal hamartoma of chest wall

Simple bone cyst

8818/0 Fibrous dysplasia

Osteofibrous dysplasia

8850/0 Lipoma NOS

8880/0 Hibernoma

Intermediate (locally aggressive)

9261/1* Osteofibrous dysplasia–like adamantinoma

8990/1 Fibrocartilaginous mesenchymoma

Malignant

9261/3 Adamantinoma of long bones

Dedifferentiated adamantinoma

8890/3 Leiomyosarcoma NOS

8802/3 Pleomorphic sarcoma, undifferentiated

Bone metastases

Haematopoietic neoplasms of bone

9731/3 Plasmacytoma of bone

9591/3 Malignant lymphoma, non-Hodgkin, NOS

9650/3 Hodgkin disease NOS

9680/3 Diffuse large B-cell lymphoma NOS

9690/3 Follicular lymphoma NOS

9699/3 Marginal zone B-cell lymphoma NOS

9702/3 T-cell lymphoma NOS

9714/3 Anaplastic large cell lymphoma NOS

9727/3 Malignant lymphoma, lymphoblastic, NOS

9687/3 Burkitt lymphoma NOS

9751/1 Langerhans cell histiocytosis NOS

9751/3 Langerhans cell histiocytosis, disseminated

9749/3 Erdheim–Chester disease

Rosai–Dorfman disease

Notes

These morphology codes are from the International Classification of Diseases for Oncology, third edition, second revision (ICD-O-3.2), available from: http://www.iacr.com.fr/index.php?option=com_content&view=article&id=149:icd-o-3-2&catid=80&Itemid=545

Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site. Behaviour code /6 is not generally used by cancer registries.

This classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.

Subtype labels are indented.

* Codes marked with an asterisk were approved by the IARC/WHO Committee for ICD-O at its meeting in January 2020.