**ICCR Soft Tissue Sarcoma Histopathology Reporting Guide – Resection Specimens, 1st edition**

**Elements in black text are CORE Elements in grey text are NON-CORE o indicates single select values □ indicates multi-select values**

|  |  |
| --- | --- |
| Definition of Core elements | Core elements are those which are essential for the clinical management, staging or prognosis of the cancer. These elements will either have evidentiary support at Level III-2 or above (based on prognostic factors in the National Health and Medical Research Council (NHMRC) levels of evidence1). In rare circumstances, where level III-2 evidence is not available an element may be made a core element where there is unanimous agreement in the expert committee. An appropriate staging system, e.g., Pathological TNM staging, would normally be included as a core element. The summation of all core elements is considered to be the minimum reporting standard for a specific cancer.**Reference** 1 Merlin T, Weston A and Tooher R (2009). Extending an evidence hierarchy to include topics other than treatment: revising the Australian 'levels of evidence'. *BMC Med Res Methodol* 9:34. |
| Definition of Non-core elements | Non-core elements are those which are unanimously agreed should be included in the dataset but are not supported by level III-2 evidence. These elements may be clinically important and recommended as good practice but are not yet validated or regularly used in patient management.Key information other than that which is essential for clinical management, staging or prognosis of the cancer such as macroscopic observations and interpretation, which are fundamental to the histological diagnosis and conclusion e.g., macroscopic tumour details, may be included as either core or non-core elements by consensus of the Dataset Authoring Committee. |
| Scope of this dataset | The dataset has been developed for the pathology reporting of resection specimens for soft tissue sarcomas. Adult rhabdomyosarcoma is also included in this dataset. A separate International Collaboration on Cancer Reporting (ICCR) dataset is available for reporting of biopsy specimens for soft tissue sarcomas.1Some soft tissue tumours which rarely arise primarily in bone and in this case should be reported using the ICCR primary tumour in bone datasets.2,3Lymphoma, uterine sarcoma, paediatric rhabdomyosarcoma and metastases are excluded from this dataset. Gastrointestinal Stromal Tumour (GIST) are also not included in this dataset as GIST displays a number of unique features which warrant its separate consideration; separate ICCR datasets for GIST are available.4,5 **References**1 International Collaboration on Cancer Reporting (2021). *Soft Tissue Sarcoma Histopathology Reporting Guide - Biopsy Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).2 International Collaboration on Cancer Reporting (2021). *Primary Tumour in Bone Histopathology Reporting Guide - Biopsy Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).3 International Collaboration on Cancer Reporting (2021). *Primary Tumour in Bone Histopathology Reporting Guide - Resection Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).4 International Collaboration on Cancer Reporting (2021). *Gastrointestinal Stromal Tumour (GIST) Histopathology Reporting Guide - Biopsy Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).5 International Collaboration on Cancer Reporting (2021). *Gastrointestinal Stromal Tumour (GIST) Histopathology Reporting Guide - Resection Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021). |

| **Core/****Non-core** | **Element name** | **Values** | **Commentary** | **Implementation notes** |
| --- | --- | --- | --- | --- |
| Non-core | CLINICAL INFORMATION | * Information not provided
* Familial syndrome, *specify*
* Multifocal disease, *specify*
* Other, *specify*
 | It is the responsibility of the clinician requesting the pathological examination of a specimen to provide information that will have an impact on the diagnostic process or affect its interpretation. The use of a standard pathology requisition/request form including a checklist of important clinical information is strongly encouraged to help ensure that important clinical data is provided by the clinician with the specimen. It is the responsibility of the pathologist to verify that all clinical information necessary for an accurate diagnosis is available to ensure that diagnosis is made within the appropriate clinical/imaging context. This can often be achieved through discussion at a multidisciplinary tumour board meeting.As an example, the coexistence of systemic disorders such as immunosuppression, which would be relevant in the evaluation of specific lesions such as Epstein-Barr virus (EBV)-related smooth muscle neoplasms and Kaposi sarcoma, should be reported. |  |
| Core | NEOADJUVANT THERAPY | * Information not provided
* Not administered
* Administered
* Neoadjuvant chemotherapy
* Neoadjuvant radiotherapy
* Other, *describe*
 | Neoadjuvant therapy may have a profound effect on the morphology of the tumour. In particular, knowledge of such prior therapy may help to interpret changes such as tumour differentiation, necrosis, vasculature changes, cellular atypia and presence of inflammatory cells. For this reason, information about any previous therapy is important for the accurate assessment of soft tissue tumour specimens.  |  |
| Core  | OPERATIVE PROCEDURE | * Not specified
* Resection, *specify if known*
* Amputation, *specify type*
* Other, *specify*
 | It is important that the type and intent of the operative procedure is clearly stated by the surgeon, as this impacts accurate pathologic assessment.  |  |
| Core | TUMOUR SITE | * Not specified
* Cutaneous, *specify deeper extension if known*
* Head and neck, specify site if known
* Trunk, *specify site and depth if known*
* Extremities, *specify site and depth if known*

 Specify laterality* Left
* Right
* Not specified
* Abdominal/pelvic visceral organ(s)*, specify site if known*
* Thoracic visceral organ(s), *specify site if known*
* Thoracic soft tissue (including mediastinum), *specify site if known*
* Retroperitoneum (including paratesticular), *specify site if known*
* Pelvis, *specify site if known*
* Other somatic or visceral site, *specify site if known*
 | Primary anatomic site is an important prognostic parameter. The anatomic location often impacts on the risk of aggressive behaviour. As an example, atypical lipomatous tumour/well differentiated liposarcoma arising superficially has a risk of local recurrence around 10%, whereas when occurring in the retroperitoneum the risk approaches 80%.  |  |
| Core  | TUMOUR DEPTH – TISSUE PLANE | * Cannot be assessed
* Not known
* Cutaneous
* Subcutaneous
* Subfascial/muscle
* Bone
* Abdominal/retroperitoneal
* Other, *specify*
 | Depth is also important. For example, the risk of distant spread of leiomyosarcoma varies from virtually 0% for purely dermal lesions to approximately 50% for deep seated tumours. For this reason, it is critical to specify anatomic location and depth as accurately as possible.The ‘not known’ designation may be necessary if tumour is excised without any surrounding normal tissue or in the absence of any information from the surgeon. |  |
| Core and Non-core | TUMOUR DIMENSIONS | Maximum tumour dimension \_\_\_ mmAdditional dimensions \_\_\_ mm x \_\_\_ mmOR* No identifiable tumour (e.g., after preoperative therapy)
* Cannot be assessed, *specify*
 | Tumour size is a critical parameter for assessment of the risk of malignant behaviour in selected histotypes such as solitary fibrous tumour.1 Size is also part of some staging systems if/when used.**Reference**1 Demicco EG, Griffin AM, Gladdy RA, Dickson BC, Ferguson PC, Swallow CJ, Wunder JS and Wang WL (2019). Comparison of published risk models for prediction of outcome in patients with extrameningeal solitary fibrous tumour. *Histopathology* 75(5):723-737. |  |
| Core | HISTOLOGICAL TUMOUR TYPE | * No residual tumour
* Atypical lipomatous tumour
* Liposarcoma, well-differentiated, *specify type*
* Dedifferentiated liposarcoma
* Myxoid liposarcoma
* Pleomorphic liposarcoma
* Dermatofibrosarcoma protuberans NOS
* Dermatofibrosarcoma protuberans, fibrosarcomatous
* Solitary fibrous tumour NOS
* Inflammatory myofibroblastic tumour
* Epithelioid inflammatory myofibroblastic sarcoma
* Myxoinflammatory fibroblastic sarcoma
* Infantile fibrosarcoma
* Fibrosarcoma NOS
* Myxofibrosarcoma
* Epithelioid myxofibrosarcoma
* Low grade fibromyxoid sarcoma
* Sclerosing epithelioid fibrosarcoma
* Plexiform fibrohistiocytic tumour
* Giant cell tumour of soft parts
* Haemangioendothelioma, *specify type*a
* Kaposi sarcoma, *specify epidemiologic type*
* Epithelioid haemangioendothelioma NOS
* Epithelioid haemangioendothelioma with *WWTR1-CAMTA1* fusion
* Epithelioid haemangioendothelioma with *YAP1-TFE3* fusion
* Angiosarcoma
* Glomus tumour, malignant
* Leiomyosarcoma NOS
* Embryonal rhabdomyosarcoma NOS
* Embryonal rhabdomyosarcoma, pleomorphic
* Alveolar rhabdomyosarcoma
* Pleomorphic rhabdomyosarcoma NOS
* Spindle cell rhabdomyosarcoma
* Osteosarcoma, extraskeletal
* Malignant peripheral nerve sheath tumour NOS
* Malignant peripheral nerve sheath tumour, epithelioid
* Malignant melanotic nerve sheath tumour
* Atypical fibroxanthoma
* Angiomatoid fibrous histiocytoma
* Ossifying fibromyxoid tumour NOS
* Synovial sarcoma, *specify type*
* Epithelioid sarcoma
* Proximal or large cell epithelioid sarcoma
* Classic epithelioid sarcoma
* Alveolar soft part sarcoma
* Clear cell sarcoma of soft tissue
* Extraskeletal myxoid chondrosarcoma
* Desmoplastic small round cell tumour
* Rhabdoid tumour of soft tissue
* Perivascular epithelioid tumour, malignant
* Myoepithelial carcinoma
* Mixed tumour, malignant, NOS
* Undifferentiated sarcoma
* Spindle cell sarcoma, undifferentiated
* Pleomorphic sarcoma, undifferentiated
* Round cell sarcoma, undifferentiated
* Ewing sarcoma
* Other round cell sarcoma, *specify*
* Sarcoma of uncertain type, *specify whether unclassifiable or requires additional testing*
* Soft tissue tumour of uncertain biologic potential, *specify type where known*
* Other, *specify*

**Diagnosis based on** (select all that apply)* Not applicable
* Morphology
* Immunohistochemistry
* Molecular testing
 | Histological diagnosis is based on the 2020 World Health Organization (WHO) Classification of Soft Tissue and Bone Tumours, 5th edition (Table 1).1 The WHO classification is based on microscopic morphologic findings, variably combined with immunohistochemical and/or molecular findings.1 If further testing is not available, then the possible diagnostic options should be described. The histopathologic report should include the supporting ancillary testing if performed.**Table 1 ( See the end of document for Tables)****References**1 WHO Classification of Tumours Editorial Board (2020). *Soft Tissue and Bone Tumours. WHO Classification of Tumours, 5th Edition, Volume 3*. IARC Publications, Lyon. 2 Fritz A, Percy C, Jack A, Shanmugaratnam K, Sobin LH, Parkin DM, Whelan SL and World Health Organization (2000). *International classification of diseases for oncology*, World Health Organization, Geneva. | This Value list based on the WHO of Soft Tissue and Bone Tumours (2020).Note that permission to publish the WHO Classification of Tumours may be needed in your implementation. It is advisable to check with the International Agency for Research on Cancer (IARC).a e.g., Kaposiform, Retiform, Pseudomyogenic, Composite or Papillary Intralymphatic angioendothelioma. |
| Core | HISTOLOGICAL TUMOUR GRADEb | * Grade 1
* Grade 2
* Grade 3
* Cannot be assessed, *specify*
 | Histologic tumour grade offers important prognostic information. While several different grading systems exist, the French1 grading system is the most widely used (see Table 2). This system is based on the assessment of differentiation, mitotic count, and necrosis.1 Importantly, the system only applies to specific histotypes (see Table 3). Many other histotypes are not gradable. Reliable tumour grading is not possible after neoadjuvant therapy.**Table 2 and 3 ( See the end of document for Tables)****Reference**1 Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, Vilain MO, Mandard AM, Le Doussal V, Leroux A, Jacquemier J, Duplay H, Sastre-Garau X and Costa J (1997). Comparative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. *J Clin Oncol* 15(1):350-362. | b Histological tumour grade is required only for specific histotypes – refer to Note, Table 3. |
| Core | MITOTIC COUNTc | \_\_\_ /2 mm2* Cannot be assessed
 | Mitotic count is a key parameter for histologic grading of malignancy as well as a factor used in risk assessment schemes (refer to **HISTOLOGICAL TUMOUR GRADE**, Table 3). The mitotic count should be determined in the most mitotic area of the tumour. The mitotic count should be reported per 2 mm2. Ten high power fields (HPFs) approximates to 2 mm2 on most modern microscopes, but the number of fields to be counted to encompass 2 mm2 should ideally be calculated on individual microscopes. | c 10 HPFs approximates to 2 mm2 on most modern microscopes, but the number of fields to be counted to encompass 2 mm2 should ideally be calculated on individual microscopes – refer to **HISTOLOGICAL TUMOUR GRADE**, Table 3. |
| Core | NECROSISd | * Not identified
* Present

 \_\_\_ % | Necrosis is a key parameter for histologic grading of malignancy. As the French grading system1 is only applicable to untreated tumours, assessment of necrosis following neoadjuvant treatment should not be performed. True coagulative necrosis (with neutrophil polymorphs and cellular debris) should be distinguished from stromal hyalinisation or infarction.**Reference**1 Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, Vilain MO, Mandard AM, Le Doussal V, Leroux A, Jacquemier J, Duplay H, Sastre-Garau X and Costa J (1997). Comparative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. *J Clin Oncol* 15(1):350-362. | d Necrosis is required for those sarcomas that are gradable – refer to **HISTOLOGICAL TUMOUR GRADE**, Table 3. |
| Non-core | LYMPHOVASCULAR INVASION | * Not identified
* Present
* Indeterminate
 | Evaluation of lymphovascular invasion has emerged as a potential prognostic parameter, however it is not yet widely adopted.1,2**References**1 Gustafson P, Akerman M, Alvegård TA, Coindre JM, Fletcher CD, Rydholm A and Willén H (2003). Prognostic information in soft tissue sarcoma using tumour size, vascular invasion and microscopic tumour necrosis-the SIN-system. *Eur J Cancer* 39(11):1568-1576.2 Engellau J, Bendahl PO, Persson A, Domanski HA, Akerman M, Gustafson P, Alvegård TA, Nilbert M and Rydholm A (2005). Improved prognostication in soft tissue sarcoma: independent information from vascular invasion, necrosis, growth pattern, and immunostaining using whole-tumor sections and tissue microarrays. *Hum Pathol* 36(9):994-1002. |  |
| Core | RESPONSE TO NEOADJUVANT THERAPY | * No prior treatment
* No response
* Response

% viable tumour \_\_\_%% necrosis \_\_\_%% therapy-induced tissue changes (e.g., fibrosis or hyalinization) \_\_\_% % of cell differentiation (e.g., myxoid liposarcoma) \_\_\_%* Cannot be assessed, *explain reasons*
 | Neoadjuvant systemic and/or local treatment of soft tissue sarcomas is gradually entering into clinical practice.1 Descriptive assessment of the amount of residual viable tumour and type of histologic response may represent valuable information in terms of estimation of efficacy of treatment. Correlation of microscopic features with macroscopic findings is critical. A scientific publication from the European Organisation for Research and Treatment of Cancer (EORTC) suggests that response should be evaluated microscopically on at least one complete central slide of tumour through its largest dimension.2 **References**1 Gronchi A, Palmerini E, Quagliuolo V, Martin Broto J, Lopez Pousa A, Grignani G, Brunello A, Blay JY, Tendero O, Diaz Beveridge R, Ferraresi V, Lugowska I, Merlo DF, Fontana V, Marchesi E, Braglia L, Donati DM, Palassini E, Bianchi G, Marrari A, Morosi C, Stacchiotti S, Bagué S, Coindre JM, Dei Tos AP, Picci P, Bruzzi P and Casali PG (2020). Neoadjuvant chemotherapy in high-risk soft tissue sarcomas: final results of a randomized trial from Italian (ISG), Spanish (GEIS), French (FSG), and Polish (PSG) Sarcoma Groups. *J Clin Oncol* 38(19):2178-2186.2 Wardelmann E, Haas RL, Bovée JV, Terrier P, Lazar A, Messiou C, LePechoux C, Hartmann W, Collin F, Fisher C, Mechtersheimer G, DeiTos AP, Stacchiotti S, Jones RL, Gronchi A and Bonvalot S (2016). Evaluation of response after neoadjuvant treatment in soft tissue sarcomas; the European Organization for Research and Treatment of Cancer-Soft Tissue and Bone Sarcoma Group (EORTC-STBSG) recommendations for pathological examination and reporting. *Eur J Cancer* 53:84-95. |  |
| Core and Non-core | MARGIN STATUS | * Cannot be assessed
* Not involved (R0)

 Distance of tumour from closest  margin \_\_\_ mm Specify closest margin, if possible Specify distance to other margin(s), if relevant* Microscopically involved (R1)

Specify margin(s), if possible* Macroscopically involved (R2)

 Specify margin(s), if possible | The status of the resection margins directly impacts patient outcome. However, there is no generally accepted way of reporting margins for soft tissue tumours. If margins are involved, a distinction is often made between microscopic involvement (R1) and resections in which it is evident macroscopically that the tumour is incompletely resected (R2). In the case of negative margins (R0), the minimum that should be documented is the distance of tumour to the closest margins. The type of tissue comprising the resection margin should also be recorded since it might be that specific tissue types (e.g., fascia) are more robust marginal tissues than others. In some cases margin status cannot be assessed for example, in liposarcomas in the retroperitoneum, or in the case of debulking, piecemeal excision or tumour rupture, in which assessment of margins is not feasible.Correlation with the surgical findings is critical to ensure accurate reporting.  |  |
| Core | LYMPH NODE STATUS | * Cannot be assessed
* No nodes submitted or found

Number of lymph nodes examined \_\_\_* Not involved
* Involved
* Number of involved lymph nodes \_\_\_
* Number cannot be determined
 | Regional lymph node metastasis is uncommon in adult soft tissue sarcomas. However, there are a few exceptions, for example epithelioid sarcoma and clear cell sarcoma of soft parts. Lymph nodes are not sampled routinely in soft tissue resections, and it is not necessary to undertake an exhaustive search for nodes. However, when present, regional lymph node metastasis has prognostic importance and should be reported.  |  |
| Non-core | COEXISTENT PATHOLOGY | * None identified
* Present
* Neoplastic pathology, *specify*
* Non-neoplastic pathology, *specify*
* Other, *specify*
 | Pathologists should report other microscopically identifiable abnormalities that are relevant to the diagnosis. For example, the presence of precursor lesions in malignant peripheral nerve sheath tumours (MPNSTs).  |  |
| Core | ANCILLARY STUDIES | * Not performed
* Performed
* Immunohistochemistry, *specify test(s) and result(s)*
* Molecular findings, *specify test(s) and result(s)*
* Other, *specify test(s) and result(s)*
 | All immunohistochemical staining and molecular tests that contributed to the diagnosis should be documented. This includes molecular testing performed on histological tumour types that are defined by specific genetic aberrations (i.e., *CIC*-rearranged sarcomas).  |  |
| Core | HISTOLOGICALLY CONFIRMED DISTANT METASTASES | * Not identified
* Present, *specify site(s)*
 | The presence of distant metastases strongly influences outcome. The pattern of metastatic spread of soft tissue sarcomas often depends on the specific histologic type. For example, metastatic spread to the lungs is very common in leiomyosarcoma whereas myxoid liposarcoma can spread to soft tissues and bone without involving the lungs.  |  |
| Non-core | PATHOLOGICAL STAGING (UICC TNM 8th edition)e | **TNM Descriptors** (only if applicable) (select all that apply) * m - multiple primary tumours
* r - recurrent
* y - post-therapy

**Primary tumour (pT)*** Inadequate specimen for assessment
* TX Primary tumour cannot be assessed
* T0 No evidence for primary tumour

EXTREMITY AND SUPERFICIAL TRUNK* T1 Tumour 5 cm or less in greatest dimension
* T2 Tumour more than 5 cm but no more than 10 cm in greatest dimension
* T3 Tumour more than 10 cm but no more than 15 cm in greatest dimension
* T4 Tumour more than 15 cm in greatest dimension

RETROPERITONEUM* T1 Tumour 5 cm or less in greatest dimension
* T2 Tumour more than 5 cm but no more than 10 cm in greatest dimension
* T3 Tumour more than 10 cm but no more than 15 cm in greatest dimension
* T4 Tumour more than 15 cm in greatest dimension

HEAD AND NECK* T1 Tumour 2 cm or less in greatest dimension
* T2 Tumour more than 2 cm but no more than 4 cm in greatest
* T3 Tumour more than 4 cm in greatest dimension
* T4a Tumour invades the orbit, skull base or dura, central compartment viscera, facial skeleton, and or pterygoid muscles
* T4b Tumour invades the brain parenchyma, encases the carotid artery, invades prevertebral muscle or involves the central nervous system by perineural spread

THORACIC AND ABDOMINAL VISCERA* T1 Tumour confined to a single organ
* T2a Tumour invades serosa or visceral peritoneum
* T2b Tumour with microscopic extension beyond the serosa
* T3 Tumour invades another organ or macroscopic extension beyond the serosa
* T4a Multifocal tumour involving no more than two sites in one organ
* T4b Multifocal tumour involving more than two sites but not more than five sites
* T4c Multifocal tumour involving more than five sites

**Regional lymph nodes (pN)*** No nodes submitted or found
* NX Regional lymph nodes cannot be assessed
* N0 No regional lymph node metastasis
* N1 Regional lymph node metastasis
 | Pathological staging is frequently not applicable or useful in most sarcoma types and has therefore been included in this dataset as a non-core element. However, staging is required in many existing reporting systems (Union for International Cancer Control (UICC)1 or American Joint Committee on Cancer (AJCC)2 8th edition staging systems), and in many cancer centres around the world it is mandated or used as a quality assurance indicator. Staging may also be required per local/institutional preference.**References**1 Brierley JD, Gospodarowicz MK and Wittekind C (eds) (2016). *Union for International Cancer Control. TNM Classification of Malignant Tumours, 8th Edition*, Wiley, USA.2 Amin MB, Edge SB, Greene FL, Byrd DR, Brookland RK, Washington MK, Gershenwald JE, Compton CC, Hess KR, Sullivan DC, Jessup JM, Brierley JD, Gaspar LE, Schilsky RL, Balch CM, Winchester DP, Asare EA, Madera M, Gress DM and Meyer LR (eds) (2017). *AJCC Cancer Staging Manual. 8th Edition*, Springer, New York. | Note that permission to publish the TNM cancer staging tables may be needed in your implementation. It is advisable to check.e Reproduced with permission.Source: UICC TNM Classification of Malignant Tumours, 8th Edition, eds by James D. Brierley, Mary K. Gospodarowicz, Christian Wittekind. 2016, Publisher Wiley (incorporating any errata published up until 6th October 2020). |

**Tables**

**Table 1: World Health Organization classification of soft tissue tumours.1**

| **Descriptor** | **ICD-O codesa** |
| --- | --- |
| **Adipocytic tumours** |  |
| *Intermediate (locally aggressive)* |  |
| Atypical lipomatous tumour | 8850/1 |
| *Malignant*   |  |
| Liposarcoma, well-differentiated, not otherwise specified (NOS) | 8851/3  |
|  Lipoma-like liposarcoma  | 8851/3 |
|  Inflammatory liposarcoma  | 8851/3 |
|  Sclerosing liposarcoma  | 8851/3 |
| Dedifferentiated liposarcoma  | 8858/3 |
| Myxoid liposarcoma  | 8852/3 |
| Pleomorphic liposarcoma | 8854/3 |
|  Epithelioid liposarcoma |  |
| Myxoid pleomorphic liposarcoma  | 8859/3\*  |
| **Fibroblastic and myofibroblastic tumours** |  |
| *Intermediate (rarely metastasizing)*  |  |
| Dermatofibrosarcoma protuberans NOS  | 8832/1 |
|  Pigmented dermatofibrosarcoma protuberans  | 8833/1  |
|  Dermatofibrosarcoma protuberans, fibrosarcomatous | 8832/3 |
|  Myxoid dermatofibrosarcoma protuberans |  |
| Dermatofibrosarcoma protuberans with myoid differentiation |  |
|  Plaque-like dermatofibrosarcoma protuberans |  |
| Solitary fibrous tumour NOS | 8815/1 |
|  Fat-forming (lipomatous) solitary fibrous tumour |  |
|  Giant cell–rich solitary fibrous tumour  |  |
| Inflammatory myofibroblastic tumour | 8825/1 |
|  Epithelioid inflammatory myofibroblastic sarcoma  |  |
| Myofibroblastic sarcoma  | 8825/3 |
| Superficial CD34-positive fibroblastic tumour  | 8810/1 |
| Myxoinflammatory fibroblastic sarcoma  | 8811/1 |
| Infantile fibrosarcoma  | 8814/3 |
| *Malignant*  |  |
| Solitary fibrous tumour, malignant  | 8815/3 |
| Fibrosarcoma NOS  | 8810/3 |
| Myxofibrosarcoma | 8811/3 |
|  Epithelioid myxofibrosarcoma  |  |
| 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  |
| **Vascular tumours** |  |
| *Intermediate (rarely metastasizing)*  |  |
| Retiform haemangioendothelioma  | 9136/1 |
| Papillary intralymphatic angioendothelioma  | 9135/1  |
| Composite haemangioendothelioma | 9136/1  |
|  Neuroendocrine composite haemangioendothelioma  |  |
| Kaposi sarcoma | 9140/3 |
|  Classic indolent Kaposi sarcoma |  |
|  Endemic African Kaposi sarcoma |  |
|  AIDS-associated Kaposi sarcoma |  |
|  latrogenic Kaposi sarcoma  |  |
| Pseudomyogenic (epithelioid sarcoma–like) haemangioendothelioma | 9138/1  |
| *Malignant*  |  |
| Epithelioid haemangioendothelioma NOS | 9133/3  |
| Epithelioid haemangioendothelioma with *WWTR1-CAMTA1* fusion |  |
| Epithelioid haemangioendothelioma with *YAP1-TFE3* fusion  |  |
| Angiosarcoma  | 9120/3 |
| **Pericytic (perivascular) tumours**  |  |
| *Malignant*   |  |
| Glomus tumour, malignant  | 8711/3  |
| **Smooth muscle tumours**  |  |
| *Malignant*   |  |
| Leiomyosarcoma NOS  | 8890/3 |
| **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  |  |
| *MYOD1*-mutant spindle cell/sclerosing rhabdomyosarcoma |  |
| Intraosseous spindle cell rhabdomyosarcoma (with *TFCP2/NCOA2* rearrangements)  |  |
| Ectomesenchymoma  | 8921/3 |
| **Chondro-osseous tumours**  |  |
| *Malignant*   |  |
| Osteosarcoma, extraskeletal  | 9180/3 |

|  |  |
| --- | --- |
| **Descriptor** | **ICD-O codesa** |
| **Peripheral nerve sheath tumours**  |  |
| *Malignant*  |  |
| Malignant peripheral nerve sheath tumour NOS  | 9540/3  |
| Malignant peripheral nerve sheath tumour, epithelioid  | 9542/3 |
| Malignant melanotic nerve sheath tumour  | 9540/3  |
| Granular cell tumour, malignant  | 9580/3  |
| **Tumours of uncertain differentiation**  |  |
| *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, NOS  | 8940/3  |
| Myoepithelioma NOS  | 8982/0 |
| *Malignant*   |  |
| Phosphaturic mesenchymal tumour, malignant NTRK-rearranged spindle cell neoplasm (emerging) | 8990/3  |
| Synovial sarcoma NOS  | 9040/3  |
| Synovial sarcoma, spindle cell  | 9041/3 |
| Synovial sarcoma, biphasic | 9043/3 |
| Synovial sarcoma, poorly differentiated  |  |
| Epithelioid sarcoma | 8804/3 |
|  Proximal or large cell epithelioid sarcoma |  |
|  Classic epithelioid sarcoma  |  |
| Alveolar soft part sarcoma  | 9581/3  |
| Clear cell sarcoma of soft tissue  | 9044/3  |
| Extraskeletal myxoid chondrosarcoma  | 9231/3  |
| Desmoplastic small round cell tumour  | 8806/3  |
| Rhabdoid tumour of soft tissue  | 8963/3  |
| Perivascular epithelioid tumour, malignant  | 8714/3  |
| Intimal sarcoma  | 9137/3 |
| Ossifying fibromyxoid tumour, malignant  | 8842/3 |
| Myoepithelial carcinoma  | 8982/3 |
| Undifferentiated sarcoma  | 8805/3 |
| Spindle cell sarcoma, undifferentiated  | 8801/3  |
| Pleomorphic sarcoma, undifferentiated  | 8802/3  |
| Round cell sarcoma, undifferentiated  | 8803/3  |
| **Undifferentiated small round cell sarcomas of bone and soft tissue** |  |
| Ewing sarcoma  | 9364/3 |
| Round cell sarcoma with *EWSR1*–non-ETS fusions  | 9366/3\*  |
| *CIC*-rearranged sarcoma  | 9367/3\*  |
| Sarcoma with *BCOR* genetic alterations  | 9368/3\*  |

a These morphology codes are from the International Classification of Diseases for Oncology, third edition, second revision (ICD-O-3.2).2 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.

\*Codes marked with an asterisk were approved by the International Agency for Research on Cancer /WHO Committee for ICD-O at its meeting in January 2020. Incorporates all relevant changes from the 5th Edition Corrigenda October 2020.

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Version 3.2 of the ICD-O codes is finalised and available at: http://www.iacr.com.fr/index.php?option=com\_content&view=article&id=149:icd-o-3-2&catid=80&Itemid=545. However, changes made to the histological entities during the 5th edition update will only be formally incorporated into a subsequent version of ICD-O once the 5th edition is complete. There are, therefore, some issues of concordance between the histological entities listed in the chapters of the WHO Classification of Tumours and the ICD-O Tables.

**Reference**

1 WHO Classification of Tumours Editorial Board (2020). *Soft Tissue and Bone Tumours. WHO Classification of Tumours, 5th Edition, Volume 3*. IARC Publications, Lyon.

**Table 2: Tumour Differentiation Score According to Histologic Type in the Updated Version of the French Federation of Cancer Centers Sarcoma Group System.1**

|  |  |
| --- | --- |
| **Histologic type** | **Score** |
| Atypical lipomatous tumour/Well-differentiated liposarcoma | 1 |
| Well-differentiated leiomyosarcoma | 1 |
| *Malignant neurofibroma* | 1 |
| *Well-differentiated fibrosarcoma* | 1 |
| Myxoid liposarcoma | 2 |
| Conventional leiomyosarcoma | 2 |
| Conventional fibrosarcoma | 2 |
| Myxofibrosarcoma | 2 |
| High-grade myxoid (round cell) liposarcoma | 3 |
| Pleomorphic liposarcoma | 3 |
| Dedifferentiated liposarcoma | 3 |
| Pleomorphic rhabdomyosarcoma | 3 |
| Poorly differentiated/pleomorphic leiomyosarcoma | 3 |
| Biphasic/monophasic/poorly differentiated Synovial sarcoma | 3 |
| Mesenchymal chondrosarcoma | 3 |
| Extraskeletal osteosarcoma | 3 |
| Extraskeletal Ewing sarcoma | 3 |
| Malignant rhabdoid tumour | 3 |
| Undifferentiated pleomorphic sarcoma | 3 |
| Undifferentiated sarcoma, not otherwise specified | 3 |

**Reference**

1 Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, Vilain MO, Mandard AM, Le Doussal V, Leroux A, Jacquemier J, Duplay H, Sastre-Garau X and Costa J (1997). Comparative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. *J Clin Oncol* 15(1):350-362.

**Table 3: Guidelines for grading soft tissue sarcomas.**

|  |  |
| --- | --- |
| **Tumours which are by definition high grade*** Ewing sarcoma
* Rhabdomyosarcoma (all types)
* Angiosarcoma
* Pleomorphic liposarcoma
* Soft tissue osteosarcoma
* Mesenchymal chondrosarcoma
* Desmoplastic small cell tumour
* Extra-renal rhabdoid tumour
* Intimal sarcoma
 | **Tumours of varying behaviour for which grading or tumour-specific risk assessment may be prognostically useful** * Myxoid liposarcoma
* Leiomyosarcoma
* Malignant peripheral nerve sheath tumour
* Solitary fibrous tumour
* Myxofibrosarcoma
* Dedifferentiated liposarcomaa
 |
| **Tumours which are by definition low grade*** Well differentiated liposarcoma/atypical lipomatous tumour
* Dermatofibrosarcoma protuberansb
* Infantile fibrosarcoma
 | **Tumours of varying behaviour for which** **grading parameters are not yet well defined*** Epithelioid hemangioendothelioma
* Extraskeletal myxoid chondrosarcoma
 |
| **Tumours which are not gradable but which often metastasize within 10-20 years of follow-up*** Alveolar soft part sarcoma
* Clear cell sarcoma
* Epithelioid sarcoma
* Synovial sarcomaa
* ‘Low-grade’ fibromyxoid sarcoma
* Sclerosing epithelioid fibrosarcoma
 |

a Some studies have shown prognostic difference between Grades 2 and 3 using the French grading system.

b Fibrosarcomatous Dermatofibrosarcoma Protuberans (DFSP) is usually regarded as intermediate grade.