**ICCR Soft Tissue Sarcoma Histopathology Reporting Guide – Biopsy 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 biopsy 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 resection specimens for soft tissue sarcomas.1  Some soft tissue tumours which rarely arise primarily in bone should be reported using the ICCR primary tumour in bone datasets.2,3  Lymphoma, 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 - Resection 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 | OPERATIVE PROCEDURE | * Not specified * Core needle biopsy * Incisional biopsy * Excisional biopsy * 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%.  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 the anatomic location and depth as accurately as possible. |  |
| 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 | Histologic 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.  Soft tissue tumours are most often first sampled by biopsy. In some cases, the biopsy is  suboptimally centred on the area(s) of interest leaving the pathologist with tissue that can be  under-representative or misrepresentative of the lesion based on the imaging studies. Molecular testing may be required to achieve a full/correct diagnosis, but the small tissue size, tissue processing issues, or suboptimal targeting of biopsy materials may make this further testing impossible. The pathologist should specify any, and all, limitations of the tissue in achieving optimal diagnosis.  **Table 1** **(See end of the 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. Even if several different systems exist, the French grading system2 is the most widely adopted (see Table 2). The system is based on the assessment of differentiation, mitotic count, and necrosis.1 Importantly, the system only applies to specific histotypes whereas many others are not gradable (see Table 3). It is important to note that grade may be underestimated in limited biopsy material.  Note: Grading sarcomas on biopsy material, using the French or any other system, can only be definitive in clearly high grade tumours. Due to the frequent morphological heterogeneity of sarcomas, high grade areas may not be included in a biopsy sample, so grading should be qualified by using phrases such as ‘at least intermediate grade’ or ‘ low grade in this limited sample’.  **Table 2 and 3** **(See end of the 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 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. |  |
| 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). |  |

**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 |
| **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.