Soft Tissue Sarcoma Histopathology Reporting Guide Biopsy Specimens



Biopsy Specimens	
Family/Last name	Date of birth DD - MM - YYYY
Given name(s)	
Patient identifiers	Date of request Accession/Laboratory number
	DD – MM – YYYY
Elements in black text are CORE. Elements in grey text ar	re NON-CORE. SCOPE OF THIS DATASET
indicates multi-select values indicates single select	
CLINICAL INFORMATION (select all that apply) (Note 1)	Abdominal/pelvic visceral organ(s), specify site if known
○ Information not provided	•
Familial syndrome, specify	
•	Thoracic visceral organ(s), specify site if known
Multifocal disease, <i>specify</i>	Thoracic soft tissue (including mediastinum), specify site if known
Training and alsease, speeny	TI KHOWII
	Retroperitoneum (including paratesticular), specify site
	▼ if known
Other, specify	
	Dolvie specify site if known
	Pelvis, specify site if known
	Other somatic or visceral site, <i>specify site if known</i>
OPERATIVE PROCEDURE (select all that apply) (Note 2)	
○ Not specified	
Core needle biopsy	
☐ Incisional biopsy	HISTOLOGICAL TUMOUR TYPE (Note 4)
Excisional biopsyOther, specify	(Value list based on the World Health Organization
Other, specify	Classification of Soft Tissue and Bone Tumours (2020))
	No residual tumour
	Atypical lipomatous tumour
	Liposarcoma, well-differentiated, specify type
TUMOUR CITE (calcab all that and b) (Note 2)	
TUMOUR SITE (select all that apply) (Note 3)	Dedifferentiated liposarcoma
Not specified	Myxoid liposarcoma
Cutaneous, specify deeper extension if known	Pleomorphic liposarcoma
	Dermatofibrosarcoma protuberans NOS
☐ Head and neck, <i>specify site if known</i>	O Dermatofibrosarcoma protuberans, fibrosarcomatous
Tread and freck, specify site if known	Solitary fibrous tumour NOS
	☐ Inflammatory myofibroblastic tumour
Trunk, specify site and depth if known	Epithelioid inflammatory myofibroblastic sarcoma
V , , , , , , , , , , , , , , , , , , ,	Myxoinflammatory fibroblastic sarcoma
	✓ Infantile fibrosarcoma✓ Fibrosarcoma NOS
Extremities, specify site and depth if known	Myxofibrosarcoma
•	Epithelioid myxofibrosarcoma
	Low grade fibromyxoid sarcoma
Specify laterality	Sclerosing epithelioid fibrosarcoma
○ Left	Plexiform fibrohistiocytic tumour
Right	Giant cell tumour of soft parts
Not specified	

Haemangioendothelioma, specify type ^a	Diagnosis based on (select all that apply)
	Not applicable
	☐ Morphology
Kaposi sarcoma, specify epidemiologic type	☐ Immunohistochemistry
7	·
	Molecular testing
Epithelioid haemangioendothelioma NOS	^a e.g., Kaposiform, Retiform, Pseudomyogenic, Composite or Papillary Intralymphatic angioendothelioma.
Epithelioid haemangioendothelioma with WWTR1- CAMTA1 fusion	h
Epithelioid haemangioendothelioma with YAP1-TFE3 fusion	HISTOLOGICAL TUMOUR GRADE ^b (Note 5) Grade 1
Angiosarcoma	Grade 2
	Grade 3
) Glomus tumour, malignant	
) Leiomyosarcoma NOS	Cannot be assessed, specify
) Embryonal rhabdomyosarcoma NOS	
Embryonal rhabdomyosarcoma, pleomorphic	
Alveolar rhabdomyosarcoma	
Pleomorphic rhabdomyosarcoma NOS	
Spindle cell rhabdomyosarcoma	^b Histological tumour grade is required only for specific histotypes – re
Osteosarcoma, extraskeletal	to Note, Table 3.
) Malignant peripheral nerve sheath tumour NOS	
	MITOTIC COUNT ^C (Note C)
Malignant peripheral nerve sheath tumour, epithelioid	MITOTIC COUNT ^c (Note 6)
Malignant melanotic nerve sheath tumour	
Atypical fibroxanthoma	/2 mm ²
Angiomatoid fibrous histiocytoma	
Ossifying fibromyxoid tumour NOS	Cannot be assessed
Synovial sarcoma, <i>specify type</i>	^c 10 HPFs approximates to 2 mm ² on most modern microscopes, but t
	number of fields to be counted to encompass 2 mm ² should ideally b
	calculated on individual microscopes – refer to Note 5, Table 3.
Epithelioid sarcoma	NECROSIS ^d (Note 7)
Proximal or large cell epithelioid sarcoma	Not identified
Classic epithelioid sarcoma	
Alveolar soft part sarcoma	Present
Clear cell sarcoma of soft tissue	%
Extraskeletal myxoid chondrosarcoma	70
Desmoplastic small round cell tumour	d Necrosis is required for those sarcomas that are gradable – refer to
	Note 5, Table 3.
Rhabdoid tumour of soft tissue	11000 57 14310 51
Perivascular epithelioid tumour, malignant	LVMPHOVACCHI AP INVACION (Note 9)
Myoepithelial carcinoma	LYMPHOVASCULAR INVASION (Note 8)
Mixed tumour, malignant, NOS	○ Not identified
Undifferentiated sarcoma	Present
Spindle cell sarcoma, undifferentiated	Indeterminate
Pleomorphic sarcoma, undifferentiated	
	COEVICIENT DATUGLOCY (Note 0)
Round cell sarcoma, undifferentiated	COEXISTENT PATHOLOGY (Note 9)
Ewing sarcoma	○ None identified
Other round cell sarcoma, specify	Present (select all that apply)
	Neoplastic pathology, <i>specify</i>
Sarcoma of uncertain type, specify whether unclassifiable or requires additional testing	
- 1. 1. 1. 1. 1. 1. 1. 1. 1. 1. 1. 1. 1.	
Soft tionus tumour of uncertain historie material	Non-neoplastic pathology, specify
Soft tissue tumour of uncertain biologic potential, specify type where known	
Other, specify	Other, specify
	V ,

Soft fissue Salconia	a biopsy Specimens
ANCILLARY STUDIES (Note 10)	
Not performedPerformed (select all that apply)	
▼	
Immunohistochemistry, specify test(s) and result(s)	
Molecular findings, specify test(s) and result(s)	
·	
Other, specify test(s) and result(s)	
•	

Definitions

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 by the Dataset Authoring Committee (DAC). 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.

Non-morphological testing e.g., molecular or immunohistochemical testing is a growing feature of cancer reporting. However, in many parts of the world this type of testing is limited by the available resources. In order to encourage the global adoption of ancillary tests for patient benefit, International Collaboration on Cancer Reporting (ICCR) recommends that some ancillary testing in ICCR Datasets is included as core elements. Where the technical capability does not yet exist, laboratories may consider temporarily using these data elements as non-core items.

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 DAC.



Scope

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 ICCR dataset is available for reporting of resection specimens for soft tissue sarcomas.²

Some soft tissue tumours which rarely arise primarily in bone should be reported using the ICCR primary tumour in bone datasets.^{3,4}

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. 5,6

The authors of this dataset can be accessed here.



Note 1 - Clinical information (Non-core)

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.



Note 2 - Operative procedure (Core)

It is important that the type and intent of the operative procedure is clearly stated by the surgeon, as this impacts accurate pathologic assessment.



Note 3 - Tumour site (Core)

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.



Note 4 - Histological tumour type (Core)

Histologic diagnosis is based on the 2020 World Health Organization (WHO) Classification of Soft Tissue and Bone Tumours, 5th edition (Table 1).⁷ The ICCR dataset includes 5th edition Corrigenda, October 2020.⁸ The WHO classification is based on microscopic morphologic findings, variably combined with immunohistochemical and/or molecular findings.⁷ 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: World Health Organization classification of soft tissue tumours.7

Descriptor	ICD-O codes ^a
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

Descriptor	ICD-O codes ^a
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 8921/3 Malignant 9180/3 Peripheral nerve sheath tumours 9540/3 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 11 Intermediate (rarely metastosizing) 8830/1 Atypical fibroxanthoma 8830/1 Angiomatoid fibrous histiocytoma 8836/1 Ossifying fibromyxoid tumour NOS 8940/0 Mixed tumour, malignant, NOS 8940/3 Myoepithelioma NOS 8982/0 Malignant 9040/3 Synovial sarcoma, spindle cell 9041/3 Synovial sarcoma, spindle cell 9041/3 Synovial sarcoma, poorly differentiated 9041/3 Epithelioid sarcoma 9581/3 Clear cell sarcoma of soft tissue 9044/3 Ex	Descriptor	ICD-O codes ^a
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		· ·
	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	· · · · · · · · · · · · · · · · · · ·	

Descriptor	ICD-O codes ^a
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).⁹ 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. Subtype labels are indented.

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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.



Note 5 - Histological tumour grade (Core)

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.¹⁰ 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'.

^{*} 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 2020.⁸

<u>Table 2: Tumour Differentiation Score According to Histologic Type in the Updated Version of the French Federation of Cancer Centers Sarcoma Group System</u>. ¹⁰

	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

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 liposarcoma
 Well differentiated liposarcoma/atypical lipomatous tumour Dermatofibrosarcoma protuberans^b 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 sarcoma^a
- 'Low-grade' fibromyxoid sarcoma
- Sclerosing epithelioid fibrosarcoma

Modified by Professor Christopher Fletcher. The original source for this information is Recommendations for the reporting of soft tissue sarcomas. Association of Directors of Anatomic and Surgical Pathology. *Mod Pathol* 1998 Dec;11(12):1257-61.¹¹



Note 6 - Mitotic count (Core)

Mitotic count is a key parameter for histologic grading of malignancy as well as a factor used in risk assessment schemes (refer to **Note 5 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 (HPF) 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.



^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.

Note 7 - Necrosis (Core)

Necrosis is a key parameter for histologic grading of malignancy. As the French grading system¹⁰ 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.

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Note 8 - Lymphovascular invasion (Non-core)

Evaluation of lymphovascular invasion has emerged as a potential prognostic parameter, however it is not yet widely adopted. 12,13

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Note 9 - Coexistent pathology (Non-core)

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).

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Note 10 - Ancillary studies (Core)

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).

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