Soft Tissue Sarcoma Classifications

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1. Introduction

We live in an age when it is increasingly important to have 'key facts' and 'headline messages'. The national registry for bone and soft tissue sarcoma want to be able to produce high level factsheets for the general public with statements such as 'There are 2000 soft tissue sarcomas annually in England' or 'Survival for soft tissue sarcomas is (eg) 75%'

It is not possible to write factsheets and data briefings like this, without a shared understanding from the SSCRG about which sarcomas we wish to include in our headline statistics.

The registry accepts that soft tissue sarcomas are a very complex and heterogeneous group of cancers which do not easily reduce to headline figures. We will still strive to collect all data from cancer registries about anything that is 'like a sarcoma'. We will also produce focussed data briefings on sites such as dermatofibrosarcomas and Kaposi's sarcomas – the aim is not to forget any sites we exclude!

The majority of soft tissue sarcomas have proved fairly uncontroversial in discussions with individual members of the SSCRG, but there were 7 particular issues it was necessary to make a group decision on. This paper records the decisions made and the rationale behind these decisions.

2. Summary of SSCRG's decisions:

Include all tumours with morphology codes as listed in Appendix A for any cancer site except C40 and C41 (bone).

Decisions:

Site	Recommendation
Kaposi's	Include
Dermatofibrosarcomas	Include
'Other' sarcomas of neither bone nor soft tissue	Include
'Not sarcomas'	Exclude
Ill defined and secondary sites	Include
Uncertain behaviour	Exclude
Morph code and site code incompatible	Include using site code, but

	improve DQ (see comments)

3. Issue by issue summary of discussions

3.1 Kaposi's Sarcomas

Relates to sarcomas with site code C46 (Kaposi's sarcoma) or morphology code M9140/3 (Kaposi's sarcoma)

Pros: Defined as sarcoma by Chas Mangham
Account for around 150 out of 2000 STS annually – significant fraction
Survival is not good – 75% at 5 years

Cons: Often described as 'not a true sarcoma as it derives from epithelial cells'
Atypical epidemiology for sarcomas linked to virus

WMCIU recommended to include
SSCRG decided to include – *Kaposi's sarcoma is a sarcoma and should be included in the headline figures.*

3.2 Dermatofibrosarcomas

Relates to sarcomas with site code C44 (non melanoma malignant neoplasm of skin)

Pros They are STS
Account for around 150 a year out of 2000 STS

Cons Cancers of skin are routinely excluded from cancer statistics by CRUK and NCIN
Dermatofibrosarcomas have very good survival (105%) so are atypical for statistics
Historically may have been coded to C49, so hard to exclude anyway, but could do it on morphology

WMCIU recommended to exclude
SSCRG decided to include – *These sarcomas may be, and arguably should be, treated in a sarcoma MDT. They count towards the workload of STS centres.*

3.3 'Other' Sarcomas of neither bone nor soft tissue

Relates to the sarcomas listed in Appendix C, which were categorised during work with Dr Mangham as neither bone nor soft tissue sarcomas but definitely true sarcomas.

Pros They are sarcomas, and should be reported somewhere in our headline figures – won't report them under bone!
They are connective tissue tumours
Would go to STS MDT usually

Cons: Advice from pathologist that they are not 'soft tissue' sarcomas.
They are mainly arising in visceral organs not regular soft tissue.

WMCIU recommended to include
SSCRG decided to include – *if we are dividing sarcomas up into bone sarcomas and soft tissue sarcomas for headline figures, they are definitely sarcomas and definitely not bone.*

3.4 Not Sarcomas

Relates to:

Mixed Mullerian Tumours M8950/3

Carcinosarcomas M8980/3, M8981/3,
Kuppfer cell sarcoma M9124/3
Gliosarcoma M9442/3
Neuroblastoma and ganglioneuroblastoma M9500.3, M9522/3 M94903
Adenosarcoma M8933/3

Pros: Sound a lot like sarcomas!
May be recorded to C49 – malignant neoplasm of soft tissue
They do have an element of sarcoma within the tumour
Some are very numerous – 70 neuroblastomas annually, 250 mixed mullerian tumours, 250 carcinosarcomas, 20 gliosarcomas, 30 adenosarcomas, 1 kuppfer cell sarcoma.

Cons: Recommended by Dr Mangham that these are not sarcomas.
Not in WHO classification as a STS

WMCIU recommended to exclude
SSCRG decided to exclude – *these sites are not true sarcomas*

3.5 Neoplasms of ill defined sites and secondary neoplasms

Relates to C76- C79. There are about 20 of these annually (out of about 2000 STS)

Pros: If a primary tumour has been registered to C76 – C79 there must be a true primary sarcoma somewhere, even if its location is unknown

Cons: Location of primary tumour is not known
Data quality around these tumours may be poor

WMCIU recommended to include, but do careful data quality on these cases.
SSCRG decided to include – *these codes do relate to a unique primary sarcoma even though the location is unknown.*

3.6 Tumours of uncertain behaviour

Relates to tumours where the behaviour code is '1', or the site code is D, not C

Pros: There are 250 annually (out of 2000 STS) – these are

Mesenchymoma (89901)	around 110 cases per year
Neurofibromatosis (95401 – included because 95403 is MPNST)	around 50 cases per year
Atypical fibrous histiocytoma (88301 – 88303 is MFH)	around 50 cases per year
Giant cell tumour of soft parts (92511)	10 cases per year
Aggressive fibromatosis (88211)	10 cases per year
Smooth muscle tumour of uncertain potential (88971)	10 cases per year

Cons: They are not included in other 'top level' cancer stats (CRUK, SEER)
We do not collect benign tumours, and so can never include these – invasive tumours is a clear line for what we report on.
They have better survival than invasive sarcomas and so may dilute messages.
Some of these conditions (notably neurofibromatosis) are no longer registrable conditions, so we would not be counting things consistently over time.

WMCIU recommended to exclude
SSCRG decided to exclude – *There was some discussion around this as some of the group were clear that these are NOT true sarcomas but can be considered to be aggressive local tumours, while others felt they should be considered as sarcomas. However, to achieve parity with national*

cancer statistics for other cancer types, and in acknowledgement that it was impossible to count benign tumours, only invasive malignancies will be counted in high level statistics.

3.7 Morphology code and site code incompatible

Relates to tumours where the morphology code and site code are incompatible, eg a chordoma of the brain rather than a chordoma of the base of skull.

Investigations suggest that 'STS' chordomas are miscodes, truly of bone, but that 'STS chondrosarcomas' do appear to be STS, down as 'extraskeletal chondrosarcomas' on path report, or in cartilage of larynx. What should be done with these?

Pros: They really are sarcomas
This includes about 50 sarcomas a year out of 2000 STS

Cons: If we do not trust the coding enough to match the morphology to a possible site, do we trust it enough to really be a sarcoma?

WMCIU recommended to include, assuming morphology correct, not site, *but* do careful data quality on these cases and try to reduce them.

WMCIU recommended further discussions specifically around extraskeletal chondrosarcomas to agree this small issue.

SSCRG decided to include, assuming site correct. *– eg if a chordoma of brain is recorded, analyse it as a soft tissue sarcoma as it is a sarcoma morphology which has been coded to a soft tissue site. If a morphology more commonly found in the soft tissue is coded to C33 or C34, analyse it as a bone sarcoma. The small number of cases mean it will not make a significant difference in analysis, and often you do get odd morphologies in strange places. However:*

SSCRG decided all cases should be sent back to registries for careful data quality check.

The SSCRG also noted that extraskeletal myxoid chondrosarcomas are a well recognised STS, and that the existence of these does not indicate a data quality problem.

Appendix A: List of codes to be included as Soft Tissue Sarcomas

ICDM	Description
87103	Glomangiosarcoma: Glomoid sarcoma
87113	Glomus tumour (and varients), malignant glomus tumour
88003	Sarcoma, NOS; soft tissue sarcoma; soft tissue tumour, malignant; mesenchymal tumour, malignant, Intimal sarcoma
88009	Sarcomatosis, NOS
88013	Spindle cell sarcoma
88033	Small cell sarcoma; round cell sarcoma
88043	Epithelioid sarcoma, epithelioid cell sarcoma
88053	Undifferentiated sarcoma
88063	Desmoplastic small round cell tumour
88103	Fibrosarcoma, NOS, sclerosing epitheliod fibrosarcoma Adult fibrosarcoma
88113	Fibromyxosarcoma Myxoinflammatory Fribroblastic sarcoma Low grade fibromyxoid sarcoma hyalinizing spindle cell tumour Myxofibrosarcoma
88133	Fascial fibrosarcoma
88143	Infantile fibrosarcoma; congenital fibrosarcoma
88153	Solitary fibrous tumour, malignant
88253	Low grade myofibroblastic sarcoma
88303	Fibrous histiocytoma, malignant; fibroxanthoma, malignant. Mailignant fibrous histiocytoma. Giant cell 'MFH' / Undifferentiated pleomorphic sarcoma with giant cells. Inflammatory 'MFH' / Undifferentiated pleomorphic sarcoma with prominent inflammation. PI
88323	Dermatofibrosarcoma, NOS (C44._); dermatofibrosarcoma protuberans, NOS (C44._)
88333	Pigmented dermatofibrosarcoma protuberans; Bednar tumour
88403	Myxosarcoma
88503	Liposarcoma, NOS; fibroliposarcoma
88513	Liposarcoma, well differentiated; Liposarcoma, differentiated Atypical lipomatous tumour lipoma-like liposarcoma
88523	Myxoid Liposarcoma; myxoliposarcoma
88533	Round cell liposarcoma
88543	Pleomorphic liposarcoma
88553	Mixed-type liposarcoma
88583	Dedifferentiated liposarcoma
88903	Leiomyosarcoma, NOS Leiomyosarcoma (excluding skin)
88913	Epithelioid leiomyosarcoma

88943	Angiomyosarcoma
88953	Myosarcoma
88963	Myxoid leiomyosarcoma
89003	Rhabdomyosarcoma, NOS; rhabdosarcoma
89013	Pleomorphic rhabdomyosarcoma
89023	Mixed type rhabdomyosarcoma
89103	Embryonal rhabdomyosarcoma; sarcoma botryoides; botryoid sarcoma. Anaplastic rhabdomyosarcoma Botryoid rhabdomyosarcoma
89123	Spindle cell rhabdomyosarcoma
89203	Alveolar rhabdomyosarcoma (incl. solid, anaplastic)
89213	Rhabdomyosarcoma with ganglionic differentiation; Ectomesenchymoma
89363	Gastrointestinal stromal sarcoma; Gastrointestinal stromal tumour, malignant; GIST malignant.
89402	Mixed tumour
89633	Rhabdoid sarcoma; Malignant rhabdoid tumour; rhabdoid tumour NOS, Extra-renal rhabdoid tumour
89903	Mesenchymoma, malignant; mixed mesenchymal sarcoma
89913	Embryonal sarcoma
90403	Synovial sarcoma, NOS; synovioma, NOS; synovioma, malignant
90413	Synovial sarcoma, spindle cell
90423	Synovial sarcoma, epithelioid cell
90433	Synovial sarcoma, biphasic
90443	Clear cell sarcoma (except of kidney M8964/3); clear cell sarcoma, of tendons and aponeuroses (C49._); melanoma, malignant, of soft parts (C49._)
91203	Haemangiosarcoma, Angiosarcoma of soft tissue Angiosarcoma
91303	Haemangioendothelioma, malignant; Hemangioendothelial sarcoma
91333	Epithelioid haemangioendothelioma, malignant
91403	Kaposi sarcoma; Multiple haemorrhagic sarcoma
91503	Haemangiopericytoma, malignant
91703	Lymphangiosarcoma; lymphangioendothelial sarcoma; lymphangioendothelioma, malignant
91803	Osteosarcoma, NOS (C40._, C41._); osteogenic sarcoma, NOS (C40._, C41._); osteochondrosarcoma (C40._, C41._); osteoblastic sarcoma (C40._, C41._) Conventional osteosarcoma Extraskeletal osteosarcoma Osteoblastic osteosarcoma Secondary osteosarcoma
92313	Myxoid chondrosarcoma, Extraskeletal Myxoid chondrosarcoma ('chordoid' type)
92513	Malignant giant cell tumour of soft parts
92523	Malignant tenosynovial giant cell tumour (C49._), giant cell tumour of tendon sheath, malignant (C49._).
92603	Ewing's sarcoma, Ewing's tumour, Extraskeletal Ewing tumour
93643	Peripheral neuroectodermal tumour PNET; neuroectodermal tumour, NOS

94733	Primitive neuroectodermal tumour, NOS: PNET, NOS: Central primitive neuroectodemal tumour, NOS (C71._): CPNET (C71._): Supratentorial PNET (C71._)
95403	Malignant peripheral nerve sheath tumour MPNST, NOS.
95603	Malignant schwannoma
95613	Malignant peripheral nerve sheath tumour with thabdomyoblastic differentiation; Triton tumour, malignant; malignant schwannoma with rhabdomyoblastic differentiation
95713	Perineurioma, malignant; Perineural MPNST
95803	Granular cell tumour, malignant; granular cell myoblastoma, malignant
95813	Alveolar soft part sarcoma
88023	Giant cell sarcoma (except of bone M9250/3); pleomorphic cell sarcoma
88573	Fibroblastic liposarcoma
88603	Angiomyoliposarcoma
89303	Endometrial stromal sarcoma (C54.1); endometrial sarcoma, NOS (C54.1); stromal sarcoma, NOS
89313	Endometrial stromal sarcoma, low grade; Endolymphatic stromal myosis; Endometrial stromatosis; Stromal endometriosis; Stromal myosis.
89353	Stromal Sarcoma
89513	Mesodermal mixed tumor
89643	Clear cell sarcoma of kidney
90203	Phyllodes tumour, malignant (C50.) Cystosarcoma phyllodes, malignant (C50.)
93653	Askin tumour
94803	Cerebellar sarcoma, NOS (C71.6) [obs]
94813	monstrocellular sarcoma C71*) ICD-O-2
95393	Meningeal sarcomatosis

Appendix B: Full list of codes discussed with decisions

ICDM	Description	Issue	Recommend
87103	Glomangiosarcoma: Glomoid sarcoma	No known issue	Include
87113	Glomus tumour (and varients), malignant glomus tumour	No known issue	Include
87131	Myopericytoma	Issue 6	Exclude
88003	Sarcoma, NOS; soft tissue sarcoma; soft tissue tumour, malignant; mesenchymal tumour, malignant, Intimal sarcoma	No known issue	Include
88009	Sarcomatosis, NOS	No known issue	Include
88013	Spindle cell sarcoma	No known issue	Include
88033	Small cell sarcoma; round cell sarcoma	No known issue	Include
88043	Epithelioid sarcoma, epithelioid cell sarcoma	No known issue	Include
88053	Undifferentiated sarcoma	No known issue	Include
88063	Desmoplastic small round cell tumour	No known issue	Include

88103	Fibrosarcoma, NOS, sclerosing epitheliod fibrosarcoma Adult fibrosarcoma	No known issue	Include
88113	Fibromyxosarcoma Myxoinflammatory Fribroblastic sarcoma Low grade fibromyxoid sarcoma hyalinizing spindle cell tumour Myxofibrosarcoma	No known issue	Include
88133	Fascial fibrosarcoma	No known issue	Include
88143	Infantile fibrosarcoma; congenital fibrosarcoma	No known issue	Include
88151	Solitary fibrous tumour, NOS	Issue 6	Exclude
88153	Solitary fibrous tumour, malignant	No known issue	Include
88211	Aggressive fibromatosis, Desmoid tumour NOS, Invasive fibroma, Extra-abdominal desmoid, Superficial fibromatoses (palmer/plantar) Desmoid-type fibromatoses	Issue 6	Exclude
88221	Abdominal fibromatosis (ICDO-2)	Issue 6	Exclude
88241	Myofibromatosis (ICD-O3)	Issue 6	Exclude
88251	Inflammatory myofibroblastic tumour, Myofibroblastic tumour, NOS	Issue 6	Exclude
88253	Low grade myofibroblastic sarcoma	No known issue	Include
88301	Atypical fibrous histiocytoma	Issue 6	Exclude
88303	Fibrous histiocytoma, malignant; fibroxanthoma, malignant. Mailignant fibrous histiocytoma. Giant cell 'MFH' / Undifferentiated pleomorphic sarcoma with giant cells. Inflammatory 'MFH' / Undifferentiated pleomorphic sarcoma with prominent inflammation. PI	No known issue	Include
88323	Dermatofibrosarcoma, NOS (C44._); dermatofibrosarcoma protuberans, NOS (C44._)	Issue 2	Include
88333	Pigmented dermatofibrosarcoma protuberans; Bednar tumour	Issue 2	Include
88341	Giant cell fibroblastoma	Issue 6	Exclude
88351	Plexiform fibrohistiocytic tumour	Issue 6	Exclude
88361	Angiomatoid fibrous histiocytoma	Issue 6	Exclude
88403	Myxosarcoma	No known issue	Include
88411	Angiomyxoma	Issue 6	Exclude
88421	Ossifying fibromyxoid tumour, atypical	Issue 6	Exclude
88501	Atypical lipoma	Issue 6	Exclude
88503	Liposarcoma, NOS; fibroliposarcoma	No known issue	Include
88513	Liposarcoma, well differentiated; Liposarcoma, differentiated Atypical lipomatous tumour lipoma-like liposarcoma	No known issue	Include

88523	Myxoid Liposarcoma; myxoliposarcoma	No known issue	Include
88533	Round cell liposarcoma	No known issue	Include
88543	Pleomorphic liposarcoma	No known issue	Include
88553	Mixed-type liposarcoma	No known issue	Include
88583	Dedifferentiated liposarcoma	No known issue	Include
88903	Leiomyosarcoma, NOS Leiomyosarcoma (excluding skin)	No known issue	Include
88913	Epithelioid leiomyosarcoma	No known issue	Include
88943	Angiomyosarcoma	No known issue	Include
88953	Myosarcoma	No known issue	Include
88963	Myxoid leiomyosarcoma	No known issue	Include
88971	Smooth muscle tumour	Issue 6	Exclude
88981	Metastasizing leiomyoma	Issue 6	Exclude
89003	Rhabdomyosarcoma, NOS; rhabdosarcoma	No known issue	Include
89013	Pleomorphic rhabdomyosarcoma	No known issue	Include
89023	Mixed type rhabdomyosarcoma	No known issue	Include
89103	Embryonal rhabdomyosarcoma; sarcoma botryoides; botryoid sarcoma. Anaplastic rhabdomyosarcoma Botryoid rhabdomyosarcoma	No known issue	Include
89123	Spindle cell rhabdomyosarcoma	No known issue	Include
89203	Alveolar rhabdomyosarcoma (incl. solid, anaplastic)	No known issue	Include
89213	Rhabdomyosarcoma with ganglionic differentiation; Ectomesenchymoma	No known issue	Include
89363	Gastrointestinal stromal sarcoma; Gastrointestinal stromal tumour, malignant; GIST malignant.	No known issue	Include
89402	Mixed tumour	No known issue	Include
89633	Rhabdoid sarcoma; Malignant rhabdoid tumour; rhabdoid tumour NOS, Extra-renal rhabdoid tumour	No known issue	Include
89821	Myoepithelioma	Issue 6	Exclude
89903	Mesenchymoma, malignant; mixed mesenchymal sarcoma	No known issue	Include
89913	Embryonal sarcoma	No known issue	Include
90403	Synovial sarcoma, NOS; synovioma, NOS; synovioma, malignant	No known issue	Include
90413	Synovial sarcoma, spindle cell	No known issue	Include
90423	Synovial sarcoma, epithelioid cell	No known issue	Include

90433	Synovial sarcoma, biphasic	No known issue	Include
90443	Clear cell sarcoma (except of kidney M8964/3); clear cell sarcoma, of tendons and aponeuroses (C49._); melanoma, malignant, of soft parts (C49._)	No known issue	Include
91203	Haemangiosarcoma, Angiosarcoma of soft tissue Angiosarcoma	No known issue	Include
91301	Haemangioendothelioma, NOS, Kaposiform haemangioepithelioma, Composite haemangioepithelioma	Issue 6	Exclude
91303	Haemangioendothelioma, malignant; Hemangioendothelial sarcoma	No known issue	Include
91331	Epithelioid haemangioendothelioma, NOS	Issue 6	Exclude
91333	Epithelioid haemangioendothelioma, malignant	No known issue	Include
91351	Endovascular papillary angioendothelioma, Retiform haemangioendothelioma, Papillary intralymphatic angioendothelioma	Issue 6	Exclude
91361	Spindle cell hemangioendothelioma	Issue 6	Exclude
91403	Kaposi sarcoma; Multiple haemorrhagic sarcoma	Issue 1	Include
91501	Haemangiopericytoma, NOS Lipomatous haemangiopericytoma	Issue 6	Exclude
91503	Haemangiopericytoma, malignant	No known issue	Include
91703	Lymphangiosarcoma; lymphangioendothelial sarcoma; lymphangioendothelioma, malignant	No known issue	Include
91741	Lymphangiomyomatosis	Issue 6	Exclude
91803	Osteosarcoma, NOS (C40._, C41._); osteogenic sarcoma, NOS (C40._, C41._); osteochondrosarcoma (C40._, C41._); osteoblastic sarcoma (C40._, C41._) Conventional osteosarcoma Extraskeletal osteosarcoma Osteoblastic osteosarcoma Secondary osteosarcoma	No known issue	Include
92313	Myxoid chondrosarcoma, Extraskeletal Myxoid chondrosarcoma ('chordoid' type)	No known issue	Include
92511	Giant cell tumour of soft parts, NOS	Issue 6	Exclude
92513	Malignant giant cell tumour of soft parts	No known issue	Include
92523	Malignant tenosynovial giant cell tumour (C49._), giant cell tumour of tendon sheath, malignant (C49._).	No known issue	Include
92603	Ewing's sarcoma, Ewing's tumour, Extraskeletal Ewing tumour	No known issue	Include
93643	Peripheral neuroectodermal tumour PNET; neuroectodermal tumour, NOS	No known issue	Include
93731	Parachondroma	Issue 6	Exclude

94733	Primitive neuroectodermal tumour, NOS: PNET, NOS: Central primitive neuroectodemal tumour, NOS (C71._): CPNET (C71._): Supratentorial PNET (C71._)	No known issue	Include
95403	Malignant peripheral nerve sheath tumour MPNST, NOS.	No known issue	Include
95603	Malignant schwannoma	No known issue	Include
95613	Malignant peripheral nerve sheath tumour with thabdomyoblastic differentiation; Triton tumour, malignant; malignant schwannoma with rhabdomyoblastic differentiation	No known issue	Include
95713	Perineurioma, malignant; Perineural MPNST	No known issue	Include
95803	Granular cell tumour, malignant; granular cell myoblastoma, malignant	No known issue	Include
95813	Alveolar soft part sarcoma	No known issue	Include
88023	Giant cell sarcoma (except of bone M9250/3); pleomorphic cell sarcoma	No known issue	Include
89333	Adenosarcoma	Issue 4	Exclude
89503	Mullerian mixed tumor	Issue 4	Exclude
89803	Carcinosarcoma NOS	Issue 4	Exclude
89813	Carcinosarcoma, embryonal	Issue 4	Exclude
91243	Kupffer cell sarcomas (C22.0)	Issue 4	Exclude
94423	Gliosarcoma (C71.): Glioblastoma with sarcomatous component (C71._)	Issue 4	Exclude
94903	Ganglioneuroblastoma	Issue 4	Exclude
95003	Neuroblastoma, NOS	Issue 4	Exclude
88271	Myofibroblastic tumor, peribronchial	Issue 6	Exclude
88573	Fibroblastic liposarcoma	Issue 3	Include
88603	Angiomyoliposarcoma	Issue 3	Include
89303	Endometrial stromal sarcoma (C54.1); endometrial sarcoma, NOS (C54.1); stromal sarcoma, NOS	Issue 3	Include
89313	Endometrial stromal sarcoma, low grade; Endolymphatic stromal myosis; Endometrial stromatosis; Stromal endometriosis; Stromal myosis.	Issue 3	Include
89353	Stromal Sarcoma	Issue 3	Include
89513	Mesodermal mixed tumor	Issue 3	Include
89643	Clear cell sarcoma of kidney	Issue 3	Include
90203	Phyllodes tumour, malignant (C50.) Cystosarcoma phyllodes, malignant (C50.)	Issue 3	Include
91611	Hemangioblastoma	Issue 6	Exclude
93653	Askin tumour	Issue 3	Include
94803	Cerebellar sarcoma, NOS (C71.6) [obs]	Issue 3	Include
94813	monstrocellular sarcoma C71*) ICD-O-2	Issue 3	Include
95393	Meningeal sarcomatosis	Issue 3	Include

Appendix C: Sarcomas of neither bone nor soft tissue

ICDM	Description	Issue	Rec.	Notes	Approx cases annually
88271	Myofibroblastic tumor, peribronchial	Issue 6	Exclude	Peribronchial. Fibromatous neoplasms	0
88573	Fibroblastic liposarcoma	Issue 3	Include	Currently not a recognised entity. Lipomatous tumours	1
88603	Angiomyoliposarcoma	Issue 3	Include	Viscera . Lipomatous tumours	1
89303	Endometrial stromal sarcoma (C54.1); endometrial sarcoma, NOS (C54.1); stromal sarcoma, NOS	Issue 3	Include	Uterus. Complex Mixed & Stromal neoplasms	80
89313	Endometrial stromal sarcoma, low grade; Endolymphatic stromal myosis; Endometrial stromatosis; Stromal endometriosis; Stromal myosis.	Issue 3	Include	Uterus. Complex Mixed & Stromal neoplasms	<5
89353	Stromal Sarcoma	Issue 3	Include	Uterus. Complex Mixed & Stromal neoplasms	<5
89513	Mesodermal mixed tumor	Issue 3	Include	Uterus. Complex Mixed & Stromal neoplasms	40
89643	Clear cell sarcoma of kidney	Issue 3	Include	Kidney. Complex Mixed & Stromal neoplasms	5
90203	Phyllodes tumour, malignant (C50.) Cystosarcoma phyllodes, malignant (C50.)	Issue 3	Include	Breast. Fibroepithelial neoplasms	40
91611	Hemangioblastoma	Issue 6	Exclude	Central Nervous System. Blood vessel tumours	<5
93653	Askin tumour	Issue 3	Include	Thoracopulmonary Ewing's sarcoma. Miscellaneous tumours	1
94803	Cerebellar sarcoma, NOS (C71.6) [obs]	Issue 3	Include	CNS. Gliomas	1
94813	monstrocellular sarcoma C71*) ICD-O-2	Issue 3	Include	CNS. Gliomas	1
95393	Meningeal sarcomatosis	Issue 3	Include	Meneinges. Meningiomas	1

Appendix D: Classifications by other organisations

Cancer Statistics (from CRUK and NCIN) for 'all cancers'

Headline figures from CRUK and NCIN report on C00-C97 excluding C44 (all invasive malignancies excluding non melanoma skin cancer). The exclude uncertain or in-situ tumours. No cancer registries collect benign tumours for sites other than brain and central nervous system.

SEER

SEER include Kaposi's Sarcoma and dermatofibrosarcomas. They exclude uncertain or in-situ tumours.

http://seer.cancer.gov/iccc/seericcc.html

Soft tissue sarcomas			
(a) Rhabdomyosarcoma and embryonal sarcoma	8900-8920, 8991	C000-C809	091
(b) Fibrosarcoma, neurofibrosarcoma and other fibromatous neoplasms	8810, 8811, 8813-8833, 9540-9561	C000-C809	092
(c) Kaposi sarcoma	9140	C000-C809	093
(d) Other specified soft-tissue sarcomas			
IX(d) excluding misclassification from III(c)	8840-8896, 8982, 8990, 9040-9044, 9120-9134, 9150-9170, 9251, 9581	C000-C809	094
	8963	C000-C639, C659-C768	094
	9231, 9240, 9363, 9364	C000-C399, C440-C809	094
	9260	C000-C399, C470-C768	094
PNET - not in brain/CNS/bone	9473 *	C000-C399,C420-C699, C730-C750,C754-C808 *	084
(e) Unspecified soft-tissue sarcomas	8800-8804	C000-C399, C440-C809	095

WHO classification of soft tissue tumours

ADIPOCYTIC TUMOURS

Benign	
Lipoma	8850/0*
Lipomatosis	8850/0
Lipomatosis of nerve	8850/0
Lipoblastoma / Lipoblastomatosis	8881/0
Angiolipoma	8861/0
Myolipoma	8890/0
Chondroid lipoma	8862/0
Extrarenal angiomyolipoma	8860/0
Extra-adrenal myelolipoma	8870/0
Spindle cell/	8857/0
Pleomorphic lipoma	8854/0
Hibernoma	8880/0
Intermediate (locally aggressive)	
Atypical lipomatous tumour/	
Well differentiated liposarcoma	8851/3
Malignant	
Dedifferentiated liposarcoma	8858/3
Myxoid liposarcoma	8852/3
Round cell liposarcoma	8853/3
Pleomorphic liposarcoma	8854/3
Mixed-type liposarcoma	8855/3
Liposarcoma, not otherwise specified	8850/3

FIBROBLASTIC / MYOFIBROBLASTIC TUMOURS

Benign	
Nodular fasciitis	
Proliferative fasciitis	
Proliferative myositis	
Myositis ossificans	
fibro-osseous pseudotumour of digits	
Ischaemic fasciitis	
Elastofibroma	8820/0
Fibrous hamartoma of infancy	
Myofibroma / Myofibromatosis	8824/0
Fibromatosis colli	
Juvenile hyaline fibromatosis	
Inclusion body fibromatosis	
Fibroma of tendon sheath	8810/0
Desmoplastic fibroblastoma	8810/0
Mammary-type myofibroblastoma	8825/0
Calcifying aponeurotic fibroma	8810/0
Angiomyofibroblastoma	8826/0
Cellular angiofibroma	9160/0
Nuchal-type fibroma	8810/0
Gardner fibroma	8810/0
Calcifying fibrous tumour	
Giant cell angiofibroma	9160/0
Intermediate (locally aggressive)	
Superficial fibromatoses (palmar / plantar)	
Desmoid-type fibromatoses	8821/1
Lipofibromatosis	
Intermediate (rarely metastasizing)	
Solitary fibrous tumour	8815/1
and haemangiopericytoma	9150/1
(incl. lipomatous haemangiopericytoma)	
Inflammatory myofibroblastic tumour	8825/1
Low grade myofibroblastic sarcoma	8825/3
Myxoinflammatory	
fibroblastic sarcoma	8811/3
Infantile fibrosarcoma	8814/3
Malignant	
Adult fibrosarcoma	8810/3
Myxofibrosarcoma	8811/3
Low grade fibromyxoid sarcoma	8811/3
hyalinizing spindle cell tumour	
Sclerosing epithelioid fibrosarcoma	8810/3

SO-CALLED FIBROHISTIOCYTIC TUMOURS

Benign	
Giant cell tumour of tendon sheath	9252/0
Diffuse-type giant cell tumour	9251/0
Deep benign fibrous histiocytoma	8830/0
Intermediate (rarely metastasizing)	
Plexiform fibrohistiocytic tumour	8835/1
Giant cell tumour of soft tissues	9251/1
Malignant	
Pleomorphic 'MFH' / Undifferentiated	
pleomorphic sarcoma	8830/3
Giant cell 'MFH' / Undifferentiated	
pleomorphic sarcoma	
with giant cells	8830/3
Inflammatory 'MFH' / Undifferentiated	
pleomorphic sarcoma with	
prominent inflammation	8830/3

* Morphology code of the International Classification of Diseases for Oncology (ICD-O) (726) and the Systematize Nomenclature of Medicine (http://snomed.org).

SMOOTH MUSCLE TUMOURS

Angioleiomyoma	8894/0
Deep leiomyoma	8890/0
Genital leiomyoma	8890/0
Leiomyosarcoma (excluding skin)	8890/3

PERICYTIC (PERIVASCULAR) TUMOURS

Glomus tumour (and variants)	8711/0
malignant glomus tumour	8711/3
Myopericytoma	8713/1

SKELETAL MUSCLE TUMOURS

Benign

Rhabdomyoma	8900/0
adult type	8904/0
fetal type	8903/0
genital type	8905/0

Malignant

Embryonal rhabdomyosarcoma	8910/3
(incl. spindle cell,	8912/3
botryoid, anaplastic)	8910/3
Alveolar rhabdomyosarcoma	
(incl. solid, anaplastic)	8920/3
Pleomorphic rhabdomyosarcoma	8901/3

VASCULAR TUMOURS

Benign

Haemangiomas of	
subcut/deep soft tissue:	9120/0
capillary	9131/0
cavernous	9121/0
arteriovenous	9123/0
venous	9122/0
intramuscular	9132/0
synovial	9120/0
Epithelioid haemangioma	9125/0
Angiomatosis	
Lymphangioma	9170/0

Intermediate (locally aggressive)

Kaposiform haemangioendothelioma	9130/1

Intermediate (rarely metastasizing)

Retiform haemangioendothelioma	9135/1
Papillary intralymphatic angioendothelioma	9135/1
Composite haemangioendothelioma	9130/1
Kaposi sarcoma	9140/3

Malignant

Epithelioid haemangioendothelioma	9133/3
Angiosarcoma of soft tissue	9120/3

CHONDRO-OSSEOUS TUMOURS

Soft tissue chondroma	9220/0
Mesenchymal chondrosarcoma	9240/3
Extraskeletal osteosarcoma	9180/3

TUMOURS OF UNCERTAIN DIFFERENTIATION

Benign

Intramuscular myxoma	8840/0
(incl. cellular variant)	
Juxta-articular myxoma	8840/0
Deep ('aggressive') angiomyxoma	8841/0
Pleomorphic hyalinizing	
angiectatic tumour	
Ectopic hamartomatous thymoma	8587/0

Intermediate (rarely metastasizing)

Angiomatoid fibrous histiocytoma	8836/1
Ossifying fibromyxoid tumour	8842/0
(incl. atypical / malignant)	
Mixed tumour/	8940/1
Myoepithelioma/	8982/1
Parachordoma	9373/1

Malignant

Synovial sarcoma	9040/3
Epithelioid sarcoma	8804/3
Alveolar soft part sarcoma	9581/3
Clear cell sarcoma of soft tissue	9044/3
Extraskeletal myxoid chondrosarcoma	9231/3
("chordoid" type)	
PNET / Extraskeletal Ewing tumour	
pPNET	9364/3
extraskeletal Ewing tumour	9260/3
Desmoplastic small round cell tumour	8806/3
Extra-renal rhabdoid tumour	8963/3
Malignant mesenchymoma	8990/3
Neoplasms with perivascular epithelioid	
cell differentiation (PEComa)	
clear cell myomelanocytic tumour	
Intimal sarcoma	8800/3