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Bone and Soft Tissue Sarcomas

Changes to Pathology Codes in the 4th Edition of the World Health Organisation Classification of Bone and Soft Tissue Sarcomas

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1.0 EXECUTIVE SUMMARY

The 4th edition of the World Health Organisation (WHO) Classification of Tumours of Soft Tissue and Bone which was published in 2012 contains notable changes from the 2002 3rd edition. The key differences between the 3rd and 4th editions can be seen in Table 1.

Table 1: Key changes to the WHO Classification of Tumours of Soft Tissue and Bone (4th edition)

ICDM Code	ICDM Handbook description	Description (third edition)	Description (fourth edition)	Status
9473	**Primitive neuroectodermal tumour**	***Discussed in WHO classification of CNS tumours***		***Obsolete***
8830	Malignant Fibrous Histiocytoma	Malignant Fibrous Histiocytoma/ undifferentiated high grade pleomorphic sarcoma	Undifferentiated high grade pleomorphic sarcoma	Current
8990	Mesenchymoma, malignant	Malignant mesenchymoma	Phosphaturic mesenchymal tumour	Current
8853	**Round cell liposarcoma**	***Round cell liposarcoma***	***Now known as myxoid liposarcoma (M8852)***	***Obsolete***
8850/3	Liposarcoma, NOS	Liposarcoma, NOS	well differentiated liposarcoma/liposarcoma NOS	Current
8850/1	Atypical lipoma		Atypical lipomatous tumour	Current
8851	Liposarcoma, well differentiated	Liposarcoma, well differentiated/ Atypical lipomatous tumour	8851/1-Lipofibromatosis (uncertain)	Current
9150	**Haemangiopericytoma**	***Haemangiopericytoma***	***Now known as solitary fibrous tumour M8815***	***Obsolete***

One of the most noticeable changes is the abolition of the term primitive neuroectodermal tumour (pNET). The term pNET was synonymous with Ewing's sarcoma in the 3rd edition of the WHO classification, often causing confusion between different groups of small round cell tumours.

The term malignant fibrous histiocytoma (MFH) has also been removed. MFH was once one of the most commonly used soft tissue sarcoma diagnoses, but the majority of tumours have been shown to be more accurately classified as a more specific sarcoma type. The small minority of tumours for which the term MFH was most representative will now be known as "undifferentiated high-grade pleomorphic sarcoma".

The term mesenchymoma has also been removed. As with MFH, it has been shown that mesenchymoma is not a distinct entity and that these sarcomas are actually a group of tumours with differing pathology. The code for mesenchymoma (8990) has been retained for 'phosphatuirc mesenchymal tumours.

A small number of tumours (approximately nine per year) were recorded as round cell liposarcomas. In the 3rd edition these were discussed along with myxoid liposarcomas. In the 4th edition, the term round cell sarcoma is a synonym for myxoid liposarcoma.

The 4th edition divides the previously synonymous terms well-differentiated liposarcoma (WDL) and atypical lipomatous tumour (ALT) into two separate categories. Though case-by-case diagnoses are left to the discretion of the clinicians involved, as a general rule tumours of this type arising in the abdominal or chest cavity or in the spermatic cord should be classified as WDL (M8850/3), tumours in the extremities are classified as ALT (M8850/1).

The term haemangiopericytoma has been removed. In the 3rd edition the term was used synonymously with the term extrapleural solitary fibrous tumour; in the 4th edition only the second term is used (M8815).

A number of new codes have been added in the 4th edition. These include gastro-intestinal stromal tumour (GIST), retiform haemangioendothelioma, and epithelioid malignant peripheral nerve sheath tumour. The addition of the GIST code provides a specific classification for tumours previously coded as mesenchymoma or leiomyosarcoma.

2.0 INTRODUCTION

Pathology techniques are continually improving, resulting in the reclassification of tumours and the identification of new morphological types and sub-types. Over the last twenty years, many new types of sarcoma have been defined, including Ewing's sarcoma and, more recently, gastro-intestinal stromal tumour (GIST). The 4th edition of the World Health Organisation (WHO) Classification of Tumours of Soft Tissue and Bone was published in 2012[1] and updates the classifications included in the 3rd edition which was published in 2002[2].

The Public Health England Knowledge and Intelligence Team (West Midlands) is the National Cancer Intelligence Network (NCIN) national lead analytical team for bone and soft tissue sarcomas. As such, it is important that any coding changes or additions in the 4th edition of the WHO classification are acknowledged and used in the analyses undertaken. This report compares the 3rd and 4th editions of "*Tumours of Soft Tissue and Bone",* and discusses the implications of key changes on reported bone and soft tissue sarcoma incidence and survival rates.

3.0 BONE AND SOFT TISSUE SARCOMA MORPHOLOGY CODES

The bone and soft tissue morphology codes to be used for the purpose of reporting bone and soft tissue sarcoma incidence and survival rates were agreed in 2011 by the NCIN's Sarcoma Site Specific Clinical Reference Group (SSCRG). The classification was based on the 3rd edition of the WHO Classification of Tumours of Soft Tissue and Bone, and is consistent with the morphology codes applied within the European Rarecare project[3] and publications reporting incidence and survival rates based on large national cancer databases[4]. It is possible for most sarcoma morphological types to arise within either bone or soft tissue (e.g. osteosarcoma, chondrosarcoma, fibrosarcoma, synovial sarcoma etc) so sarcoma morphologies predominantly arising within bone were not distinguished from those of soft tissue, and vice versa. A table summarising omissions, additions and changes in terminology between the 3rd and 4th editions of the WHO Classification of Tumours of Soft Tissue and Bone is included in Appendix A.

4.0 KEY CHANGES

The most significant changes in the 4th edition of the WHO sarcoma classification are as follows:

I. Peripheral Neuroectodermal Tumours (pNET)

ICDM Code:	**M9364/M9473**
Annual Incidence:	**24 tumours (excluding pNETs of the brain)**
2002 terminology:	**Peripheral/primitive neuroectodermal tumour (pNET)**
2012 terminology:	**Extra-skeletal Ewing's sarcoma (M9364)**

Ewing's sarcoma and pNET were discussed as a single entity within the 3rd edition, and defined as round cell sarcomas which show "varying degrees of neuroectodermal differentiation"[5]. pNET tumours were said to belong to the "Ewing family of tumours" (term now also obsolete) and were often used interchangeably with Ewing's sarcoma. This terminology caused much confusion, as pNET refers to a small cell sarcoma and the term could apply to non-sarcomatous tumours such as medulloblastoma[6], which do not possess the Ewing's sarcoma translocation t(11;22). The removal of the term pNET from the new WHO classification should eradicate any further confusion distinguishing between different types of small cell tumours.

Between 1990 and 2010, there were 717 diagnoses of soft tissue Ewing's sarcomas and pNETs in the UK. Of these, 299 were Ewing's sarcomas, 320 were primitive neuroectodermal tumours and 98 were peripheral neuroectodermal tumours (excluding sarcomas of the brain). The morphology code for primitive neuroectodermal tumour is present in the 4th edition, although the term has now been changed to "extra-skeletal Ewing's sarcoma". Thus, any tumours diagnosed with this morphology code should possess the t(11; 22) translocation, though this may not be true of all cases.

The removal of pNETs from Ewing's sarcoma analyses means that Ewing's sarcoma age-standardised incidence rates (ASRs) decrease significantly (Figure 1a). The addition of peripheral neuroectodermal tumours which possess the t(11;22) translocation associated with Ewing's sarcomas, which have been reclassified as extra-skeletal Ewing's sarcomas[7] has little counterbalancing effect due to the very small numbers. The omission of pNETs has a positive effect on Ewing's sarcoma survival rates but the change is not statistically significant (Figure 1b).

Figure 1a: Ewing's sarcoma 3-year rolling age-standardised incidence rates (UK: 1990-2010)

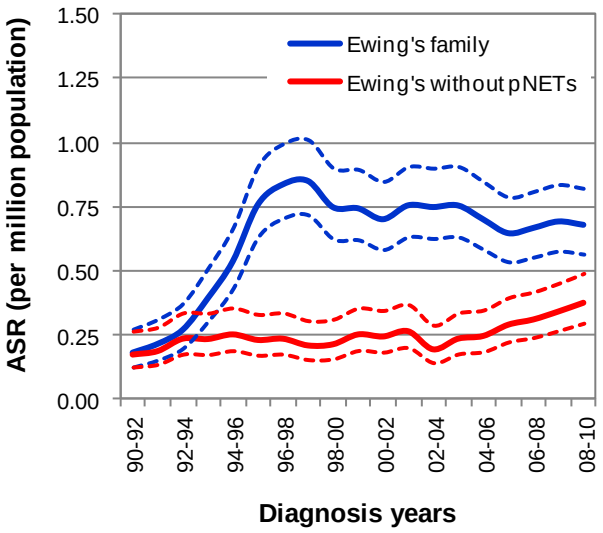
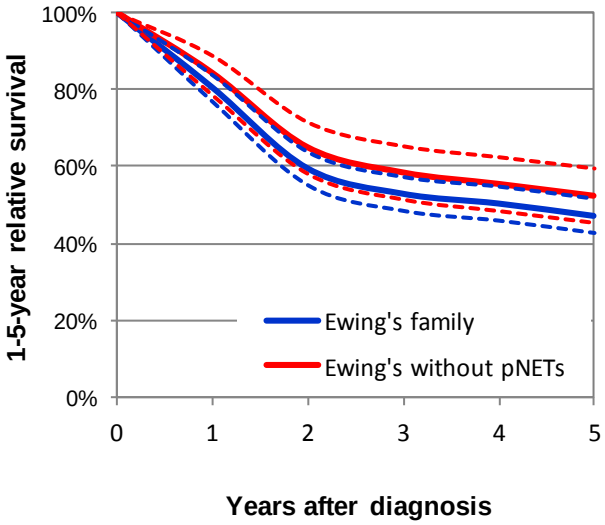


Figure 1b: Ewing's sarcoma 5-year relative survival rates (UK: 1990-2005)



II. Malignant Fibrous Histiocytoma

ICDM Code: **M8830**
Annual Incidence: **233 tumours in 1990, decreasing to 92 tumours in 2010**
2002 terminology: **Malignant Fibrous Histiocytoma**
2012 terminology: **Undifferentiated high-grade pleomorphic sarcoma (M8830)**

Historically, the term MFH was applied to pleomorphic spindle cell neoplasms with fibroblastic and histiocytic differentiation. However, the majority of tumours within this classification did not show any evidence of histiocytotic differentiation, and many were found to be other types of poorly differentiated tumours. Hence, the term MFH is now only applicable to a small subset of tumours known as "pleomorphic sarcomas with no differentiation".

Malignant fibrous histiocytoma (M8830) was once amongst the most commonly diagnosed variant of soft tissue sarcoma, and the annual incidence has gradually diminished, to 109 diagnoses in 2009 and 92 in 2010. The term MFH is now obsolete and it has been replaced by the term "Undifferentiated high-grade pleomorphic sarcoma". These changes will have the greatest impact on incidence and survival rates pertaining to "fibroblastic" sarcomas.

III. Mesenchymoma

ICDM Code: **M8990**
Annual Incidence: **Unknown as GISTs were coded to this ICDM.**
2002 terminology: **Mesenchymoma**
2012 terminology: ***Obsolete but code retained for "phosphaturic mesenchymal tumour"***

Mesenchymoma – (M8990) once applied synonymously with the term GIST, the term "mesenchymoma" has been removed from the soft tissue sarcoma classification. The true incidence of this tumour was never known for certain as the allocated morphology code included

GISTs, although mesenchymomas are known to have been very rare. It has also been shown that tumours that would previously have been categorised as mesenchymomas are generally more accurately classified as other tumours[8]. A GIST specific morphology code was introduced in ICD-03, and with all cancer registries now coding in ICD-03, the true incidence of phosphaturic mesenchymal tumours should be distinguishable.

IV. Round Cell Liposarcoma

ICDM Code:	**M8853**
Annual Incidence:	**9 per year**
2002 terminology:	**Round Cell Liposarcoma**
2012 terminology:	**Myxoid Liposarcoma (M8852)**

Round cell sarcomas were discussed as an entity along with "myxoid liposarcomas" within the 3rd edition, and described as "lesions formally known as round cell liposarcoma".

Action: Tumours diagnosed in recent years as round cell liposarcomas will be treated as myxoid liposarcomas for the purpose of analysis.However, it is important to note that within this myxoid liposarcoma code there will likely be a range of tumours behaving differently dependent on their round cell component, though distinguishing between these will be very difficult.

V. Well-differentiated liposarcomas (WDL)/Atypical Lipomatous Tumour (ALT)

ICDM Code:	**M8851/1**	**Atypical lipomatous tumour**
ICDM Code:	**M8851/3**	**Well differentiated liposarcoma**
Annual Incidence (extremities):		**83 per year**
2002 terminology:	**ALT/WDL**	
2012 terminology:	**ALT**	**(M8850/1)**
	WDL	**(M8850/3)**

In the 3rd edition, atypical lipomatous tumours (ALTs) and well differentiated liposarcoma (WLDs) are discussed as a single entity. Within the 4th edition, a greater emphasis is placed on the term ALT. If a well-differentiated liposarcoma arises within the abdominal / pelvic cavity, such as the retroperitoneum or mediastinum, it is difficult to obtain wide margins, and these tumours frequently recur locally. They also have a greater propensity to dedifferentiate into high grade sarcomas and to ultimately result in death. However, if an ALT/WLD arises within the extremities, then the tumours are curable through simple excision, and thus, tend not to recur. Therefore, it is questionable whether the terminology sarcoma is applicable to a WDL arising in the extremities, and the term ALT may be more appropriate. The 4th edition states that the underlying classification for these tumours is the decision of the responsible surgeon and pathologist. Inconsistencies between surgeons and pathologists in different organisations may therefore lead to spurious geographical variations in the incidence and survival rates for these tumours in national analyses.

The change in coding will pose problems itself. WDLs were originally coded to M8851, but the 4th edition specifies that the morphology code M8850 should be used. This is the code used for liposarcoma, not otherwise specified (NOS). Thus, if this new code is introduced for WDLs, it will not be possible to distinguish between WDLs and miscoded liposarcoma tumours in the future.

Hypothetically, if all extremity WDLs were diagnosed as ALTs, then the incidence of WDLs would be significantly lower (Figure 2a) as would the overall incidence of liposarcomas (Figure 2b). When examining the impact of the exclusion of extremity WDLs on the incidence rates of all extremity sarcomas, it is possible to see a small but consistent and statistically significant reduction in incidence (Figure 2c). 5-year relative survival is not significantly reduced by the exclusion of all extremity WDLs in patients diagnosed in 1990-1994 and 1997-2001, but for patients diagnosed in 1998-2002 and 2001-2005, 5-year relative survival is reduced significantly (Figure 2d).

Figure 2a: Well-differentiated liposarcoma 3-year rolling age standardised incidence rates (UK: 1990-2010)

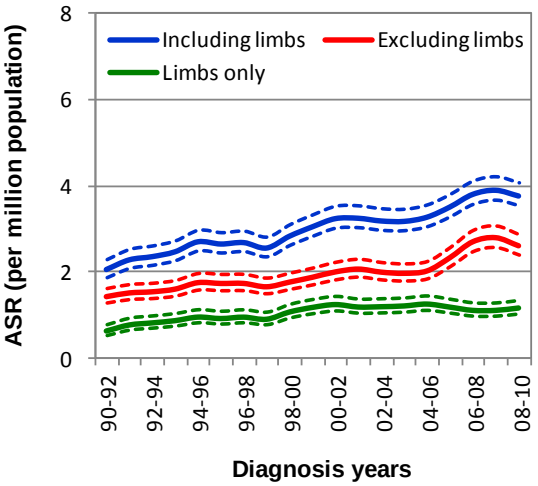


Figure 2b: Liposarcoma 3-year rolling age-standardised incidence rates (UK: 1990-2010)

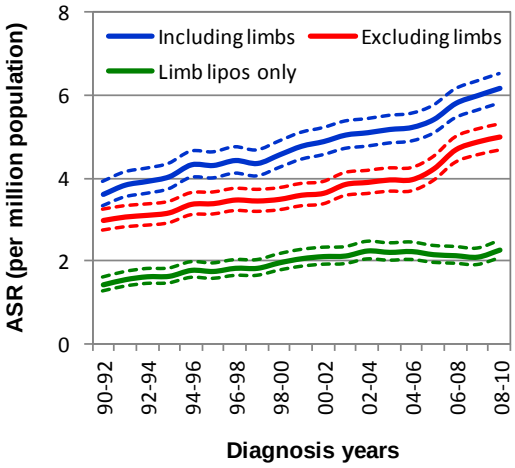


Figure 2c: Extremity sarcoma 3-year rolling age standardised incidence rates (UK: 1990-2010)

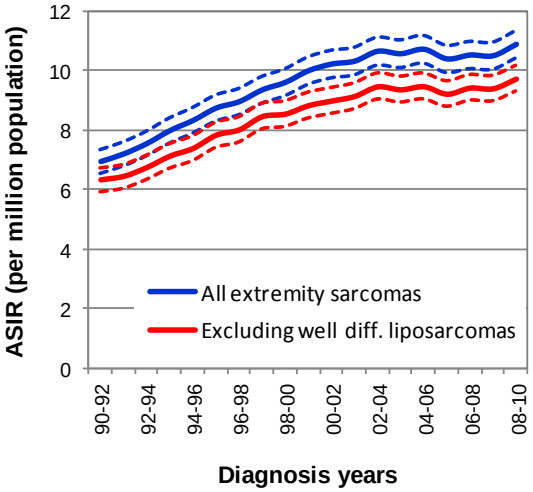
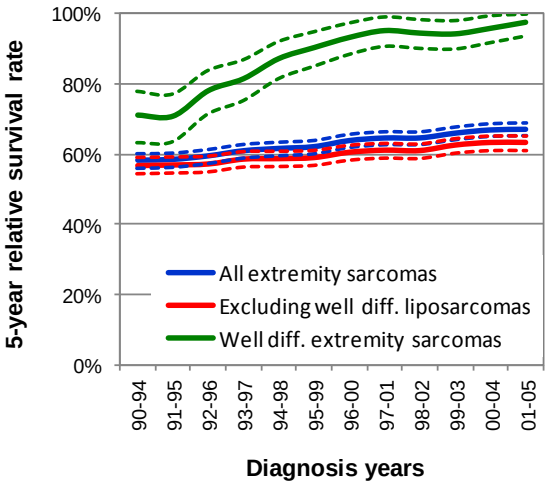


Figure 2d: 5-year relative survival of extremity sarcomas including and excluding extremity WDLs (UK: 1990-2005)



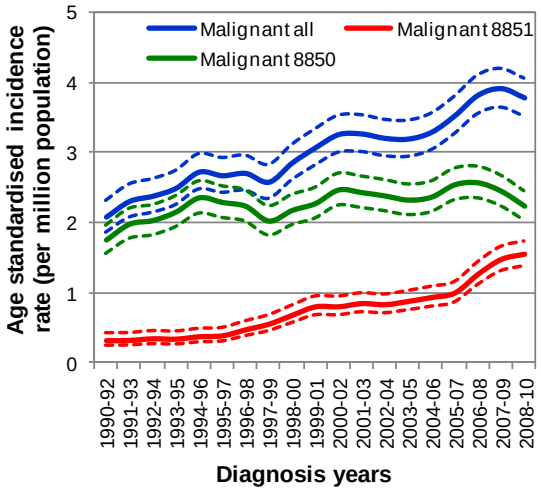


Figure 2e: Incidence of liposarcoma NOS, WDL and the effect of reclassifying all WDLs to M8850

If well-differentiated liposarcomas (currently M8851) are to be recoded as recommended in the 4th edition to the same morphology code as liposarcoma NOS (M8850), the overall incidence of liposarcoma NOS will increase significantly (Figure 2e). Moreover, in the future, it will not be possible to distinguish between liposarcoma NOS and well-differentiated liposarcoma.

Action: Guidance will be required from the sarcoma SSCRG to discuss whether extremity WDLs should be included in sarcoma incidence and survival statistics. It may be most useful to always differentiate between limb and abdominal cavity tumours as a matter of course, and always refer to them separately while calculating incidence and survival.

VI. Haemangiopericytoma

ICDM Code:	**M9150**
Annual Incidence:	**11 per year**
2002 terminology:	**Haemangiopericytoma**
2012 terminology:	**Solitary fibrous tumour (M8815)**

Haemangiopericytoma is described alongside "extrapleural solitary fibrous tumour" in the 3rd edition, and it was observed that distinguishing between the two terms had become arduous. The term haemangiopericytoma has thus been defined as obsolete, and these tumours will now be referred to as "solitary fibrous tumour". This will have little or no impact on the underlying sarcoma incidence and survival rates as only the terminology has changed.

Action: For future analyses, all haemangiopericytomas diagnosed in the past can be recoded to M8815 so a complete incidence pattern for this tumour type may be established.

VII. Obsolete terms with <5 diagnoses annually

Embryonal sarcoma M8991 (Incidence 2008-2010: 8 tumours)
Synonymous with embryonal rhabdomyosarcoma

Myosarcoma M8895 (Incidence 2008-2010: 8 tumours)
Synonymous with embryonal rhabdomyosarcoma

Mixed Liposarcoma M8855 (Incidence 2008-2010: 5 tumours)
Obsolete – possibly described by alternative liposarcoma term

Lymphangiosarcoma M9170 (Incidence 2008-2010: 2 tumours)
This term is an amalgamation of both cancer site "lymph" and sarcoma type angiosarcoma.
Hence, synonymous with angiosarcoma.

Parachordoma M9373 (Incidence 2008-2010: 0 tumours)
Usually a tumour of uncertain malignant potential

Intra-osseous low grade M9187 (Incidence 2008-2010: 0 tumours)
Osteosarcoma Possibly a synonym for "low grade central osteosarcoma"

Synovial sarcoma, M9042 (Incidence 2008-2010: 0 tumours)
epithelioid cell Obsolete

Action: To ensure these changes are reflected in future analyses, all instances of the old morphology codes will be updated and allocated to the new morphology codes.

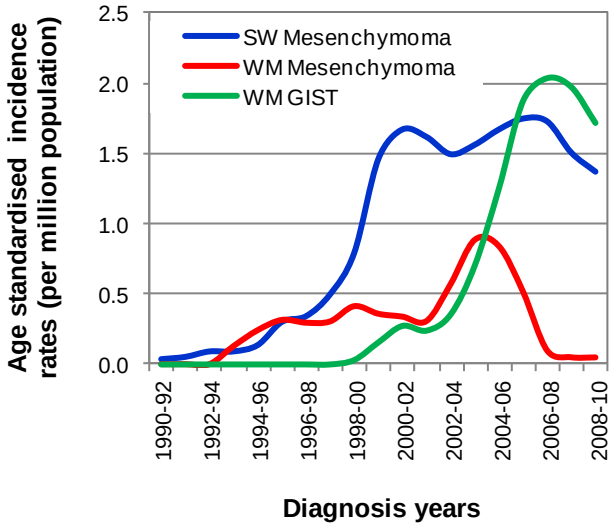
VIII. New ICDM Codes

PEComa	M8714
Overlap with other WHO classifications[9] (although discussed in the 3rd edition, no allocated morphology code was identified and underlying tumours were discussed in the WHO classification of renal, hepatic, and pulmonary tumours).	
Retiform Haemangioendothelioma	M9137
Pseudomyogenic Haemangioendothelioma	M9138
Epithelioid Malignant Peripheral Nerve Sheath Tumour	M9542
Gastro-intestinal stromal tumour	M8936 (present in ICD-03 so currently in use)
Phosphuratic mesenchymal tumour	M8990

IX. Gastro-intestinal stromal tumours

Gastro-intestinal stromal tumours were not described in the 3rd edition, and were previously classified as mesenchymomas, and before that as leiomyosarcomas. Some cancer registries have been applying the specific GIST ICD-O3 morphology code since 2006. This creates considerable inconsistencies in GIST incidence rates across regions, and makes calculating the true incidence of GIST very challenging. The figure below compares incidence rates of mesenchymoma and GIST in two cancer registry regions; the West Midlands (WM), where the specific GIST code has been in use since 2006, and the South West (SW), where the specific GIST morphology code as not yet been applied. As the number of GISTs recorded increases significantly in the West Midlands, the number of recorded mesenchymomas approaches zero. In contrast, the number of mesenchymomas in the South West remains stable for much of the latter half of the time period, before falling slightly in the later years.

Figure 3: GIST and mesenchymoma 3-year rolling age standardised incidence rates (WM and SW: 1990-2010)



5.0 IMPACT ON INCIDENCE AND SURVIVAL RATES

The changes in incidence and survival resulting from individual amendments within the 4th edition were discussed in the previous sections. Figures 3a and 3b show the effect of the omission of extremity ALTs and pNETs on overall soft tissue sarcoma incidence and survival rates.

Using the new WHO sarcoma classification overall soft tissue sarcoma incidence rates are marginally, although not statistically significantly lower (Figure 3a). These amendments have no impact on 5-year relative survival rates (Figure 3b). The changes in the 4th edition only affect a fraction of the sarcomas diagnosed annually. These analyses are purely hypoethetical as "pNETs" with the t(11; 22) translocation will still be classified as Ewing's sarcomas.

Figure 3a: Incidence rate of all soft tissue sarcoma including and excluding extremity ALTs and pNETs (UK: 1990-2010)

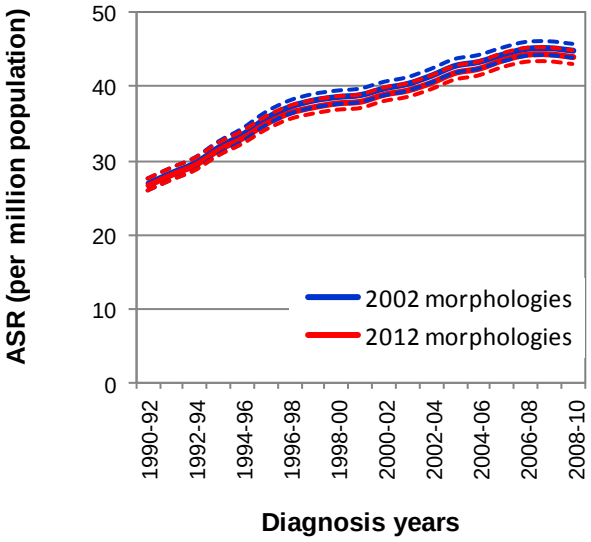
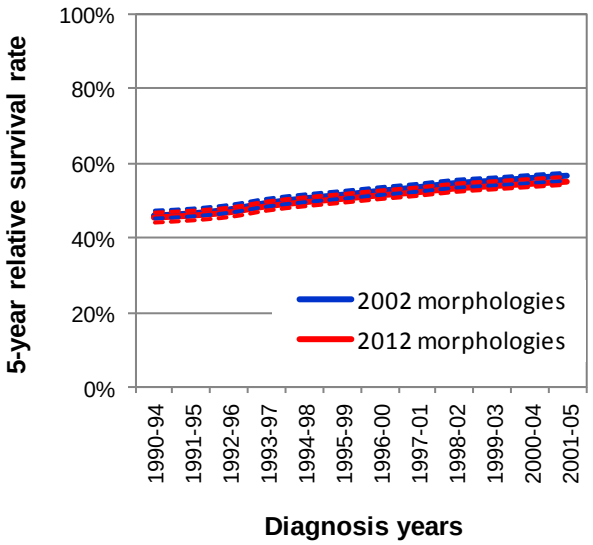


Figure 3b: 5-year relative survival of all soft tissue sarcomas including and excluding extremity ALTs and pNETs (UK: 1990-2005)



DISCUSSION

Although a small proportion of the WHO bone and soft tissue sarcoma terms are now obsolete, and new concepts have been introduced, this does not necessarily mean that pathologists will immediately stop using the old terminology. Thus, certainly for the immediate future, terms such as "Fibromyxosarcoma" may still arise, instead of the new WHO terminology "Myxoinflammatory fibroblastic sarcoma". However, with the release of the new classification, terminology such as "peripheral neuroectodermal tumour" has been eradicated, and these tumours will be labelled with a more specific round cell sarcoma description.

The new classification does not remove inconsistencies in the diagnosis and reporting of well-differentiated liposarcomas of the extremities. Five-year relative survival rates for these tumours are close to 100% as well-differentiated sarcomas in the extremities are curable through wide resection, and hence, applying the term "sarcoma" to these tumours may not be warranted. However, the 4th edition states that the decision to code an extremity sarcoma as an atypical lipomatous tumour or a well-differentiated liposarcoma is open to the responsible surgeon and/or pathologist.

The 4th edition describes GISTs for the first time. Trying to understand the incidence and survival patterns of these tumours has presented enormous challenges. Historically, GISTs were misclassified as leiomyosarcomas, and later as mesenchymomas. Thus, the true incidence of GISTs (as well as leiomyosarcomas) may not be ascertained for another two or three years, and it may be many more years before national survival data can be obtained.

For future analyses of bone and soft tissue sarcomas, obsolete terms and ICDM codes which are no longer in use will still be applied, but the new ICDM codes identified within the 4th edition will be incorporated whenever possible. In instances where certain tumour types have been found to be synonymous with other sarcomas, a recoding exercise will be undertaken so that the sarcomas are included with the updated terminology.

Appendix A- Counts of diagnoses in England

2008-2010 incidence	ICDM Code	ICDM Handbook description	Description (third edition)	Description (fourth edition)	Status
0	8710	Glomangiosarcoma	Glomangiosarcoma	Glomangiosarcoma (also known as malignant glomus tumour [M8711])	Current
1	8711	Malignant glomus tumour	Malignant glomus tumour	Malignant glomus tumour	Current
0	8713	Glomangiomyoma	Myopericytoma	Glomangiomyoma (also known as malignant glomus tumour [M8711])	Current
-	8714	-	*new term*	PEComa NOS, malignant	Current
993	8800	Sarcoma, NOS	Intimal sarcoma	Not discussed	Current
464	8801	Spindle cell sarcoma	Not discussed	Undifferentiated spindle cell sarcoma	Current
228	8802	Giant cell sarcoma	Not discussed	Undifferentiated pleomorphic sarcoma	Current
38	8803	Small cell sarcoma	Not discussed	Undifferentiated round cell sarcoma	Current
67	8804	Epithelioid sarcoma	Epithelioid sarcoma	Undifferentiated epithelioid sarcoma	Current
0	8805	Undifferentiated sarcoma	Not discussed	Undifferentiated sarcoma, NOS	Current
0	8806	Desmoplastic small round cell tumour	Desmoplastic small round cell tumour	Desmoplastic small round cell tumour	Current
155	8810	Fibrosarcoma, NOS	Adult fibrosarcoma/sclerosing epithelioid fibrosarcoma	Adult fibrosarcoma	Current
360	8811	Fibromyxosarcoma	Myxofibrosarcoma/Myxoinflammatory fibroblastic sarcoma	Myxofibrosarcoma	Current
1	**8812**	**Periosteal fibrosarcoma**	***not included***	***not included***	***Obsolete***
0	8813	Fascial fibrosarcoma	Not discussed	Palmar/plantar fibromatosis	Current
9	8814	Infantile fibrosarcoma	Infantile fibrosarcoma	Infantile fibrosarcoma	Current
0	8815	Solitary fibrous tumour, NOS	Solitary fibrous tumour	Solitary fibrous tumour	Current
0	8821	Aggressive fibromatosis	Superficial fibromatoses/desmoid-type fibromatoses	Desmoid-type fibromatosis	Current
0	8822	Abdominal fibromatosis	Not discussed	Abdominal fibromatosis	Current
1	8823	Desmoplastic fibroma	Desmoplastic fibroma of bone	Desmoplastic fibroma of bone	Current
0	8824	Myofibromatosis	Myofibromatosis	Myofibromatosis (benign)	Current
0	8825	Myofibroblastic tumour, NOS	Low-grade myofibroblastic sarcoma	Low-grade myofibroblastic sarcoma	Current
230	8830	Malignant Fibrous Histiocytoma	Malignant Fibrous Histiocytoma/ undifferentiated high grade pleomorphic sarcoma	Undifferentiated high grade pleomorphic sarcoma	Current
423	8832	Dermatofibrosarcoma, NOS	***Described in the WHO classification of skin tumours***	Dermatofibrosarcoma	Current
24	8833	Pigmented dermatofibrosarcoma		Pigmented dermatofibrosarcoma	Current

2008-2010 incidence	ICDM Code	ICDM Handbook description	Description (third edition)	Description (fourth edition)	Status
0	8834	Giant cell fibroblastoma	Not discussed	Giant cell fibroblastoma	Current
0	8835	Plexiform fibrohistiocytic tumour	Plexiform fibrohistiocytic tumour	Plexiform fibrohistiocytic tumour	Current
0	8836	Angiomatoid fibrous histiocytoma	Angiomatoid fibrous histiocytoma	Angiomatoid fibrous histiocytoma	Current
63	8840	Myxosarcoma	Intramuscular myxoma (benign)	Low-grade fibromyxoid sarcoma/sclerosing epithelioid fibrosarcoma	Current
2	8841	Angiomyxoma	Angiomyxoma	Angiomyxoma (benign)	Current
0	8842	Ossifying fibromyxoid tumour	Ossifying fibromyxoid tumour	Ossifying fibromyxoid tumour	Current
415	8850/3	Liposarcoma, NOS	Liposarcoma, NOS	well differentiated liposarcoma/liposarcoma NOS	Current
	8850/1			Atypical lipomatous tumour	Current
301	8851	Liposarcoma, well differentiated	Liposarcoma, well differentiated/ Atypical lipomatous tumour	8851/1-Lipofibromatosis (uncertain)	Current
189	8852	Myxoid Liposarcoma	Myxoid Liposarcoma	Myxoid Liposarcoma	Current
17	**8853**	**Round cell liposarcoma**	***Round cell liposarcoma***	***Now known as myxoid liposarcoma (M8852)***	***Obsolete***
77	8854	Pleomorphic liposarcoma	Pleomorphic liposarcoma	Pleomorphic liposarcoma	Current
5	**8855**	**Mixed liposarcoma**	***Mixed liposarcoma***	***not included***	***Obsolete***
0	8857	Fibroblastic liposarcoma	Spindle cell lipoma (benign)	Spindle cell lipoma (benign)	Current
152	8858	Dedifferentiated liposarcoma	Dedifferentiated liposarcoma	Dedifferentiated liposarcoma	Current
1	8860	Angiomyoliposarcoma	***Discussed in WHO classification of Urogenital tumours***	Extra-renal angiomyolipoma (benign)	Current
1484	8890	Leiomyosarcoma, NOS	Leiomyosarcoma	Leiomyosarcoma	Current
31	8891	Epithelioid leiomyosarcoma	***Discussed in Who classification of breast and female genital organs***		Current
21	8894	Angiomyoma	Angioleiomyoma, angionyoma (usually benign)	Angioleiomyoma, angionyoma (usually benign)	Current
7	**8895**	**Myosarcoma**	***Now known as embronal rhabdomyosarcoma (M8910)***		***Obsolete***
21	8896	Myxoid leiomyosarcoma	***Discussed in Who classification of breast and female genital organs***		Current
1	8897	Smooth muscle tumour	***Discussed in Who classification of breast and female genital organs***		Current
0	8898	Metastising leiomyosarcoma	***Discussed in Who classification of breast and female genital organs***		Current
112	8900	Rhabdomyosarcoma, NOS	Rhabdomyoma	Rhabdomyoma	Current
31	8901	Pleomorphic rhabdomyosarcoma	Pleomorphic rhabdomyosarcoma	Pleomorphic rhabdomyosarcoma	Current
3	**8902**	**Mixed type rhabdomyosarcoma**	***Synonym for alveolar rhabdomyosarcoma?????????????***		***Obsolete?***
83	8910	Embryonal rhabdomyosarcoma	Embryonal rhabdomyosarcoma	Embryonal rhabdomyosarcoma	Current
0	8912	Spindle cell rhabdomyosarcoma	Spindle cell rhabdomyosarcoma	Spindle cell/sclerosing rhabdomyosarcoma	Current

2008-2010 incidence	ICDM Code	ICDM Handbook description	Description (third edition)	Description (fourth edition)	Status
72	8920	Alveolar rhabdomyosarcoma	Alveolar rhabdomyosarcoma	Alveolar rhabdomyosarcoma	Current
0	8921	Rhabdomyosarcoma with ganglionic differentiation	Not discussed	Ectomesenchymoma	Current
174	8930	Endometrial stromal sarcoma	***Discussed in Who classification of breast and female genital organs***		Current
9	8931	Endometrial stromal sarcoma	***Discussed in Who classification of breast and female genital organs***		Current
0	**8935**	**Stromal Sarcoma**	***Not discussed***	***Not discussed***	***Obsolete***
0	8936	Gastrointestinal stromal sarcoma	***not discussed***		Current
49	8963	Rhabdoid sarcoma	***Extra-renal rhabdoid tumour***		Current
14	8964	Clear cell sarcoma of kidney	***Discussed in WHO Classification of Urinary and Male Genital Organs***		Current
21	8982	Myoepithelioma	Myoepithelioma	Myoepithelial carcinoma	Current
478	8990	Mesenchymoma, malignant	Malignant mesenchymoma	Phosphaturic mesenchymal tumour	Current
8	**8991**	**Embryonal sarcoma**	***Synonym for embryonal rhabdomyosarcoma M8910***		***Obsolete?***
150	9020	Phyllodes tumour, malignant	**Discussed in Who classification of breast and female genital organs**		Current
167	9040	Synovial sarcoma, NOS	Synovial sarcoma	Synovial sarcoma	Current
26	9041	Synovial sarcoma, spindle cell	Synovial sarcoma, spindle cell	Synovial sarcoma, spindle cell	Current
0	**9042**	**Synovial sarcoma, epithelioid cell**	***Not discussed***	***Not discussed***	***Obsolete***
16	9043	Synovial sarcoma, biphasic	Synovial sarcoma, biphasic	Synovial sarcoma, biphasic	Current
32	9044	Clear cell sarcoma	Clear cell sarcoma of soft tissue	Clear cell sarcoma of soft tissue	Current
333	9120	Haemangiosarcoma	Angiosarcoma of soft tissue	Angiosarcoma of soft tissue	Current
12	9130	Haemangioendothelioma	Kaposiform/Composite haemangioendothelioma	Kaposiform/Composite haemangioendothelioma	Current
22	9133	Epithelioid haemangioendothelioma	Epithelioid haemangioendothelioma	Epithelioid haemangioendothelioma	Current
0	9135	Endovascular papillary angioendothelioma	Retiform/papillary intralymphatic haemangioendothelioma	Papillary intralymphatic angioendothelioma	Current
0	9136	Spindle cell hemangioendothelioma	Not discussed	Retiform/composite haemangioendothelioma	Current
-	9137		new term	Intimal sarcoma	Current
-	9138		new term	Pseudomyogenic haemangioendothelioma	Current
427	9140	Kaposi sarcoma	Kaposi sarcoma	Kaposi sarcoma	Current
30	**9150**	**Haemangiopericytoma**	***Haemangiopericytoma***	***Now known as solitary fibrous tumour M8815***	***Obsolete***
2	**9170**	**Lymphangiosarcoma**	***Lymphangioma (benign)***	***Synonym for angiosarcoma M9120***	***Obsolete***
0	**9174**	**Lymphangiomyomatosis**	***not included***	***not included***	***Obsolete***

2008-2010 incidence	ICDM Code	ICDM Handbook description	Description (third edition)	Description (fourth edition)	Status
359	9180	Osteosarcoma, NOS	Osteosarcoma	Osteosarcoma	Current
36	9181	Chondroblastic osteosarcoma	Chondroblastic osteosarcoma	Chondroblastic osteosarcoma	Current
9	9182	Fibroblastic osteosarcoma	Fibroblastic osteosarcoma	Fibroblastic osteosarcoma	Current
12	9183	Telangiectatic osteosarcoma	Telangiectatic osteosarcoma	Telangiectatic osteosarcoma	Current
7	**9184**	**Osteosarcoma in Paget's disease of bone**	***Paget osteosarcoma***	***Not discussed as a separate entity***	***Obsolete?***
1	9185	Small cell osteosarcoma	Small cell osteosarcoma	Small cell osteosarcoma	Current
0	**9186**	**Central osteosarcoma**	***Now known as low-grade central osteosarcoma (M9187)***		***Obsolete***
0	9187	Intraosseous well differentiated osteosarcoma	Low-grade central osteosarcoma	Low-grade central osteosarcoma	Current
4	**9190**	**Juxtacortical osteosarcoma**	***Synonym of parosteal osteosarcoma***	***Now known as Parosteal osteosarcoma (M9192)***	***Obsolete***
0	9192	Parosteal osteosarcoma	Parosteal osteosarcoma	Parosteal osteosarcoma	Current
0	9193	Periosteal osteogenic sarcoma	Periosteal sarcoma	Periosteal sarcoma	Current
0	9194	High grade surface osteosarcoma	High grade surface osteosarcoma	High grade surface osteosarcoma	Current
0	**9195**	**Intracortical osteosarcoma**	***Intracortical osteosarcoma***	***Not discussed***	***Obsolete***
1	9200	Aggressive osteoblastoma	Aggressive osteoblastoma	Aggressive osteoblastoma	Current
1	9210	Osteochondromatosis	Osteochondromatosis	Osteochondromatosis	Current
480	9220	Chondrosarcoma	Chondrosarcoma	Chondrosarcoma	Current
3	9221	Juxtacortical chondrosarcoma	Periosteal chondrosarcoma	Periosteal chondrosarcoma	Current
3	9230	Chondroblastoma, malignant	Chondroblastoma	Chondroblastoma	Current
46	9231	Myxoid chondrosarcoma	Myxoid chondrosarcoma	Myxoid chondrosarcoma	Current
10	9240	Mesenchymal chondrosarcoma	Mesenchymal chondrosarcoma	Mesenchymal chondrosarcoma	Current
0	9242	Clear cell chondrosarcoma	Clear cell chondrosarcoma	Clear cell chondrosarcoma	Current
0	9243	Dedifferentiated chondrosarcoma	Dedifferentiated chondrosarcoma	Dedifferentiated chondrosarcoma	Current
29	9250	Giant cell tumour of bone	Giant cell tumour of bone	Giant cell tumour of bone	Current
7	9251	Giant cell tumour of soft parts	Giant cell tumour of soft tissues	Giant cell tumour of soft tissues	Current
0	9252	Malignant tenosynovial giant cell tumour	Giant cell tumour of tendon sheath	Malignant tenosynovial giant cell tumour	Current
260	9260	Ewing's sarcoma	Ewing sarcoma	Ewing sarcoma	Current
3	9261	Adamantinoma of long bones	Adamantinoma	Adamantinoma	Current
9	9270	Odontogenic tumour	***Discussed in WHO classification of Head and Neck Tumours***		Current

2008-2010 incidence	ICDM Code	ICDM Handbook description	Description (third edition)	Description (fourth edition)	Status
0	9290	Ameloblastic odontosarcoma	***Discussed in WHO classification of Head and Neck Tumours***		Current
7	9310	Ameloblastoma	***Discussed in WHO classification of Head and Neck Tumours***		Current
1	9330	Ameloblastic fibrosarcom	***Discussed in WHO classification of Head and Neck Tumours***		Current
0	9341	Clear cell odontogenic tumour	***Discussed in WHO classification of Head and Neck Tumours***		Current
0	9342	Odontogenic carcinomsarcoma	***Discussed in WHO classification of Head and Neck Tumours***		Current
40	9364	Peripheral neuroectodermal tumour	Primitive neuroectodermal tumour	Extraskeletal Ewing sarcoma	Current
0	**9365**	**Askin tumour**	***Askin tumour***	***Synonym for Ewing's sarcoma arising in the chest wall***	***Obsolete***
122	9370	Chordoma	Chordoma	Chordoma	Current
0	9371	Chondroid chordoma	Chondroid chordoma	Chondroid chordoma	Current
0	9372	Dedifferentiated chordoma	Dedifferentiated chordoma	Dedifferentiated chordoma	Current
0	**9373**	**Parachordoma**	***Parachordoma***	***Synonym for myoepothelioma M8982***	***Obsolete***
78	**9473**	**Primitive neuroectodermal tumour**	***Discussed in WHO classification of CNS tumours***		***Obsolete***
168	9540	Malignant peripheral nerve sheath tumour MPNST, NOS	Not discussed	Malignant peripheral nerve sheath tumour	Current
-	9542		new term	Epithelioid malignant peripheral nerve sheath tumour	Current
51	9560	Neurilemoma, malignant	Neurilemoma	Neurilemoma	Current
9	9561	Malignant peripheral nerve sheath tumour with Rhabdomyoblastic differentiation	Not discussed	Malignant triton tumour	Current
0	9571	Perineurioma	Not discussed	Perineurioma	Current
5	9580	Granular cell tumour, malignant	Not discussed	Malignant granular cell tumour	Current

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