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Bone and Soft Tissue Sarcomas

UK Incidence and Survival:
1996 to 2010

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Version 2.0

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Version 2.0

This is an updated version of the UK Incidence and Survival 1996 to 2010 report published in July 2013.

Alterations made to the first version

- Sarcomas of the skin diagnosed in Scotland are included
- Skin sarcomas are excluded from survival rates in all UK countries

CONTENTS

EXECUTIVE SUMMARY

Primary bone and soft tissue sarcomas are an exceptionally rare form of cancer, collectively accounting for only 1% of all malignancies diagnosed. This report fills a void in publicly available data describing the incidence and survival of patients diagnosed with primary bone or soft tissue sarcoma in the UK. The report considers overall UK incidence and survival rates, and examines variations amongst the four UK countries. Differences between UK countries in the most common histologies and anatomical sites are also discussed.

In 2010 there were 531 new diagnoses of bone sarcoma and 3,298 new diagnoses of soft tissue sarcoma in the UK. The age-standardised incidence of bone sarcoma remained constant at around 7.9 per million between 1996 and 2010. Soft tissue sarcoma incidence rates increased significantly from 39 per million to 45 per million during the same time period. This increase may reflect improved diagnostic techniques and reporting rather than a true increase in incidence.

The incidence of soft tissue sarcomas increased significantly with increasing age. The age specific incidence rate was highest in males aged 85 years and over where it reached 230 per million and exceeded the rate for females by a ratio of 1.9:1. Age specific incidence rates for soft tissue sarcomas in females aged 45 to 59 years were slightly higher than those in males, due to the incidence of gynaecological sarcomas. Age specific incidence rates for bone sarcoma were bi-modal, with peaks in incidence observed in teenagers and adolescents, as well as the elderly. Bone sarcoma age specific incidence rates were significantly higher in males than females over 15 years of age. Rates in males exceeded those of females by a ratio of 1.7:1 in those aged 15 to 19 years and by a ratio of 1.6:1 in those aged 55 years and over.

The most common site for both bone and soft tissue sarcomas was within the extremities (23% of soft tissue sarcomas and 52% of bone sarcomas). The proportion of Kaposi's sarcoma was significantly higher in England due to high levels of HIV-linked Kaposi's sarcoma in the Thames region. Scotland and Wales had a significantly higher proportion of soft tissue sarcomas arising within the gastro-intestinal tract. Owing to the coding practice used within the Northern Ireland Cancer Registry, Northern Ireland had a significantly higher proportion of tumours diagnosed within the thorax and trunk and a significantly lower proportion within the retroperitoneum. This coding practice is the same that is used for retroperitoneal sarcomas by some of the sarcoma specialist centres in England. A national consensus relating to the coding of retroperitoneal sarcomas needs to be agreed if meaningful data on the management of these tumours is to be made available to support centralised specialised commissioning in England.

Leiomyosarcoma (22%) and liposarcoma (12%) were the most common soft tissue sarcoma sub-types diagnosed in the UK. Dermatofibrosarcomas accounted for 5% of sarcomas in England, 4% in Wales, 7% in Northern Ireland and 3.5% in Scotland. Northern Ireland had a higher proportion of rhabdomyosarcomas, which could be explained by the younger population age structure within Northern Ireland, and a significantly lower proportion of myxofibrosarcomas (<1%) compared to the UK average (3%). The latter could be a reflection of current pathological reporting within Northern Ireland and requires further investigation.

There were significant improvements in soft tissue sarcoma 5-year relative survival rates over the 10-year period studied, with rates increasing from 51% in 1996-2000 to 55% in 2006-2010. From the year 2000 onwards, 5-year relative survival was significantly higher in males than females, by a difference of up to four percentage points. This could be due to the increased incidence of well-differentiated liposarcomas in males. Bone sarcoma 5-year relative survival rates did not change significantly in the 10-year period studied and increased by only two percentage points from 54% in 1996-2000 to 56% in 2001-2005. There were no significant differences between the 5-year relative survival rates for bone sarcomas diagnosed in males and females.

1.0 INTRODUCTION

Bone and soft tissue sarcomas are a group of rare heterogeneous forms of cancer, which collectively account for approximately 1% of all malignancies diagnosed. Sarcomas represent a challenge to clinicians as they are rare and diagnosis is often delayed. Best practice guidelines advise that they should be treated within centres that specialise in the management of sarcomas. There is a void in the published literature documenting the incidence and survival of bone and soft tissue sarcomas in the UK, as most existing publications are based on either hospital databases or cohort studies. This report presents incidence and survival data for all patients diagnosed with malignant bone or soft tissue sarcomas in the UK between 1996 and 2010.

There are over one hundred different morphological sub-types of sarcoma (Appendix A). The most common types of bone sarcoma are osteosarcoma, chondrosarcoma, Ewing's sarcoma and chordoma. Soft tissue sarcomas develop from soft tissue cells including smooth muscle cells (leiomyosarcomas), fat cells (liposarcomas), fibrous connective tissue (fibrosarcomas), skeletal muscles (rhabdomyosarcomas), synovium (synovial sarcomas), blood vessels (angiosarcomas), breast ducts (phyllodes tumours) and nerves (nerve sheath tumours). Kaposi's sarcoma is a particular type of sarcoma linked to immune deficiency in patients with HIV infection and is included in this report for completeness. Gastro-intestinal stromal tumours (GISTs) occur in the stomach or small intestine and were frequently coded as leiomyosarcomas in the past. As there are inconsistencies in the coding of GISTs across the United Kingdom (UK), incidence and survival data have not been included as a separate entity in this report, and these tumours have been included in the leiomyosarcoma category.

This report presents data for the 5 most common variants of bone sarcoma. The 22 most common variants of soft tissue sarcoma have been grouped into 12 sub-types, and the remaining less common variants are represented in a separate group. For the purpose of reporting incidence and survival, soft tissue sarcomas in the 12 morphological sub-type groups were further combined according to the anatomical diagnostic site groupings in Appendix B.

2.0 METHODS

The Public Health England, Knowledge and Intelligence Team (West Midlands) (WM KIT) is the National Cancer Intelligence Network (NCIN) lead intelligence team in England for bone and soft tissue sarcoma. As such, the WM KIT analyses national data on the incidence, mortality, survival and treatment of bone and soft tissue sarcomas in England. These analyses are usually conducted using the National Cancer Data Repository (NCDR), a compilation of data collected by the eight English regional cancer registration offices that now form the National Cancer Registration Service (NCRS). The current version of the NCDR also includes cancer registry data from Scotland, Wales and Northern Ireland, enabling the analyses of bone and soft tissue sarcomas within the UK.

Bone and soft tissue sarcomas are classified by both the tenth revision of the International Classification of Diseases (ICD-10) site code and their morphology. ICD-10 is the standard global system for reporting mortality and morbidity statistics[1]. Within that system, the prefix 'C' locates the code within the 'neoplasm', or cancer, sub-group, and the numbers that follow localise the tumour to a specific area of the body. A two number string denotes a general area of the body, while a three number string represents a more specific area; for example, 'C-03' denotes a malignant neoplasm of the gum, and 'C-031' represents a malignant neoplasm of the lower gum. Bone sarcomas are identified specifically from ICD-10 site codes C40 (malignant neoplasm of bone and articular cartilage of limbs) and C41 (malignant neoplasm of bone and articular cartilage of other and unspecified sites). Sarcomas arising within all anatomical cancer sites outside of the bone are soft tissue sarcomas.

All malignant sarcomas (excluding sarcomas of the brain, ICD-10 codes C70, C71 and C72) are included in the analyses in this report. The current version of the NCDR includes all malignancies

diagnosed in England in the period 1990 to 2010. However, data for Northern Ireland do not commence until 1993. Potential coding issues with regard to the soft tissue sarcomas recorded within the NCDR also exist, particularly those registered in the early 1990's. Therefore, the analyses in this report have been restricted to incidence and survival rates for bone and soft tissue sarcomas diagnosed in the UK in the 15-year period between 1996 and 2010.

Confidence intervals around incidence rates were calculated using the gamma method. Relative survival is defined as the observed survival in the patient group divided by the expected survival of the general population, matched by age, sex, and calendar year. Relative survival was calculated in Stata (v.11) using the strs programme which calculates relative survival estimates using the Ederer II method[2]. National life tables were obtained from the Cancer Research UK Cancer Survival Group at the London School of Hygiene and Tropical Medicine. Five-year relative survival was calculated using 5-year rolling averages.

3.0 DATA QUALITY

Historically, to construct the NCDR, the eight NCRS cancer registration offices in England submitted the details for all malignant tumours recorded within their local cancer registration database, regardless of whether or not the patient resided within the corresponding cancer registry's catchment area. Thus, cancer registries submitted both intra- and extra-regional tumour details. In the most recent iteration of the NCDR covering tumour diagnoses between 1990 and 2010, the cancer registration offices were asked to submit only their intra-regional cases in order to avoid duplication of tumour details.

An investigation of the NCDR was undertaken to identify the number of potential duplicate sarcomas recorded in the latest iteration of the NCDR. Three hundred and fourteen (approximately 14 to 15 per year) sarcomas appear to have been recorded more than once either within the same registry, or across two or more cancer registries. The number of potential duplicate tumours recorded annually is insignificant in comparison to the 2,800 sarcomas diagnosed annually. However, the greatest issue appears to relate to 1999 where the tumour details for around 50 sarcomas appear to have been duplicated. There are also "pockets" of the country where the United Kingdom and Ireland Association of Cancer Registries (UKIACR) postcode directory specifies that the postcode belongs to a particular cancer registry, but where two (or more) registration offices have submitted cancer registration details for the same sarcoma.

Table 1: Potential duplication of tumours recorded within the NCDR (England: 1990-2010)

Cancer Registration Office	East Midlands	Eastern Office	North West	Northern & Yorkshire	South East	South West	Thames	West Midlands	Grand Total
East Midlands	23			2					**25**
Eastern Office	2	35		2					**39**
North West	2	1	11	2	1	9	8		**34**
Northern & Yorkshire				17					**17**
South East	2	1			5		5		**13**
South West	8	8		4		70	19		**109**
Thames	5	7		3			53		**68**
West Midlands	2			1		1	1	4	**9**
Grand Total	**44**	**52**	**11**	**31**	**6**	**80**	**86**	**4**	**314**

Table 1 summarises the duplicate sarcomas identified. Around 70% of the duplicate registrations were within the same cancer registration office (i.e. a single cancer registration office has registered the same tumour more than once). The remaining duplicates were cross-regional (i.e. more than one registration office has registered the same tumour). The potential duplicates identified in Table 1 are to be queried directly with the appropriate cancer registration office.

4.0 INCIDENCE

In the UK between 1996 and 2010, 43,590 patients were diagnosed with a soft tissue sarcoma (3,298 in 2010) and 7,229 patients with a bone sarcoma (531 in 2010). In 2010, the soft tissue

sarcoma age-standardised incidence rate (ASR) was 45 per million, and the bone sarcoma age-standardised incidence rate (ASR) was 8.2 per million (Figures 1a and 1b). These observed incidence rates are similar to the incidence rates for bone and soft tissue sarcoma reported by other cancer registries outside the UK[3,4,5].

Figure 1a: Five-year rolling soft tissue sarcoma age-standardised incidence rates (UK: 1996-2010)

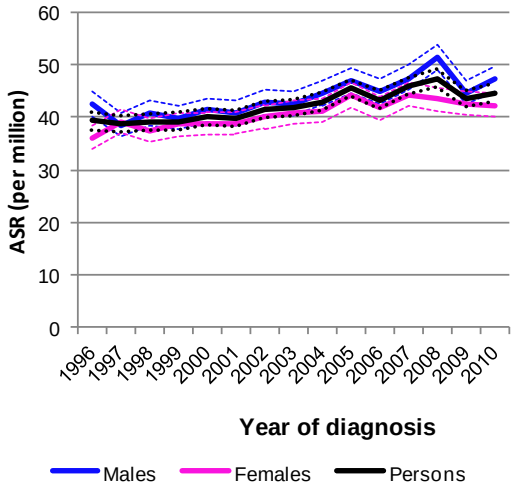
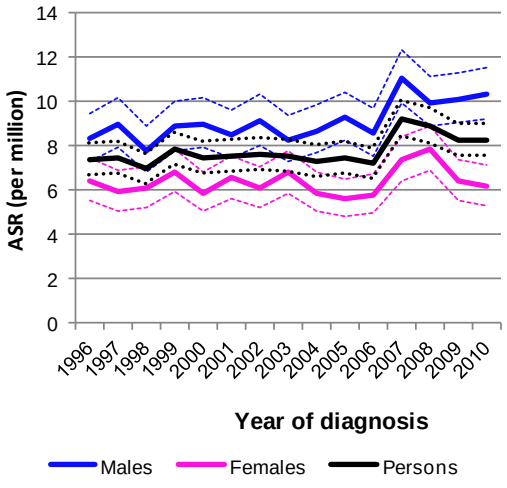


Figure 1b: Five-year rolling bone sarcoma age-standardised incidence rates (UK: 1996-2010)



Soft tissue sarcoma age-standardised incidence rates have increased gradually, but significantly, from 39 per million in 1996 to 45 per million in 2010. Bone sarcoma incidence rates remained constant between 1996 and 2006 at 7.4 per million, but increased significantly to 9.2 per million in 2007. Between 2009 and 2010, bone sarcoma incidence rates decreased slightly to 8.2 per million, and are not significantly higher than the incidence rate observed between 1996 and 2006. The higher bone sarcoma incidence rates in 2007 and 2008 were confined to England and were predominantly in the Thames Cancer Registry catchment area.

4.1 Variation in Bone and Soft Tissue Sarcoma Incidence Rates with Sex

Male soft tissue sarcoma age-standardised incidence rates were higher than those in females throughout the 15-year period studied, but the rates were significantly higher only in 1996, 2008 and 2010 (Figure 1a). Bone sarcoma age-standardised rates in males were also consistently higher than those of females, and this difference was statistically significant between 2004 and 2010 (Figure 1b).

4.2 Variation in Bone and Soft Tissue Sarcoma Incidence Rates with Age

The incidence of soft tissue sarcomas increased significantly with increasing age (Figure 2a). The age specific incidence rate (ASIR) was highest in males aged 85 years and over where it reached 230 per million (23 per 100,000) and exceeded the rate for females by a ratio of 1.9:1. Age specific incidence rates for soft tissue sarcomas in females aged 45 to 59 years were slightly higher than those in males due to the incidence of gynaecological sarcomas.

Age specific incidence rates (ASIR) for bone sarcoma were bi-modal, with peaks in incidence observed in teenagers and adolescents, as well as the elderly (Figure 2b). Bone sarcoma age specific incidence rates were significantly higher in males than in females over 15 years of age. Rates in males exceeded those of females by a ratio of 1.7:1 in those aged 15 to 19 years and by a ratio of 1.6:1 in those aged 55 years and over.

Figure 2a: Soft tissue sarcoma age specific incidence rates (United Kingdom: 1996-2010)

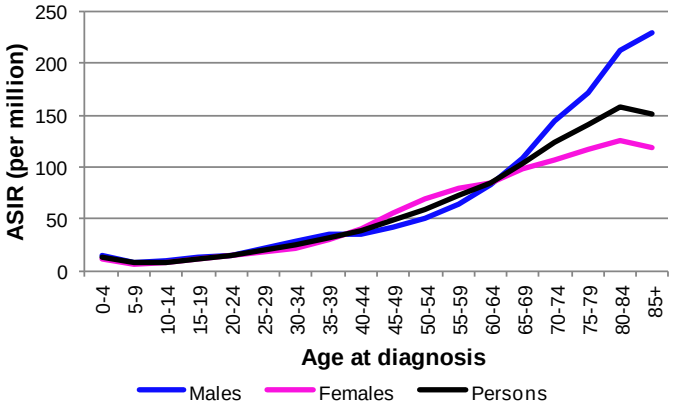
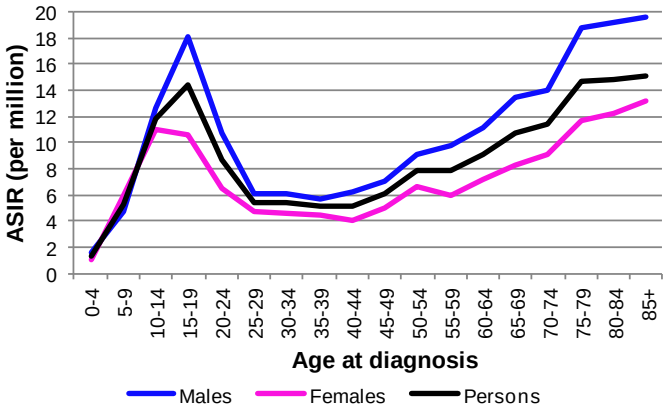


Figure 2b: Bone sarcoma age specific incidence rates (United Kingdom: 1996-2010)



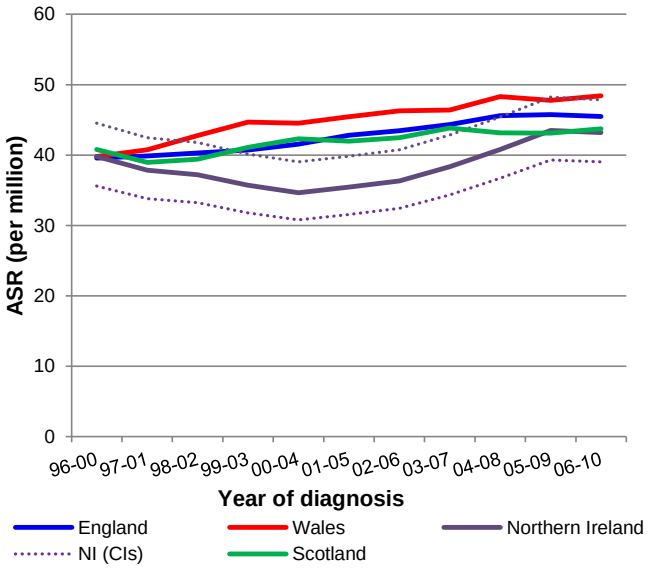
4.3 Variation in Bone and Soft Tissue Sarcoma Incidence Rates with UK Country

On average, 2,740 soft tissue sarcomas were diagnosed annually in England, 270 in Scotland, 180 in Wales and 80 in Northern Ireland. In 2010, 443 bone sarcomas were diagnosed in England, 48 in Scotland, 23 in Wales and 17 in Northern Ireland (Table 2).

Table 2: United Kingdom populations and bone and soft tissue sarcoma incidence (2010)

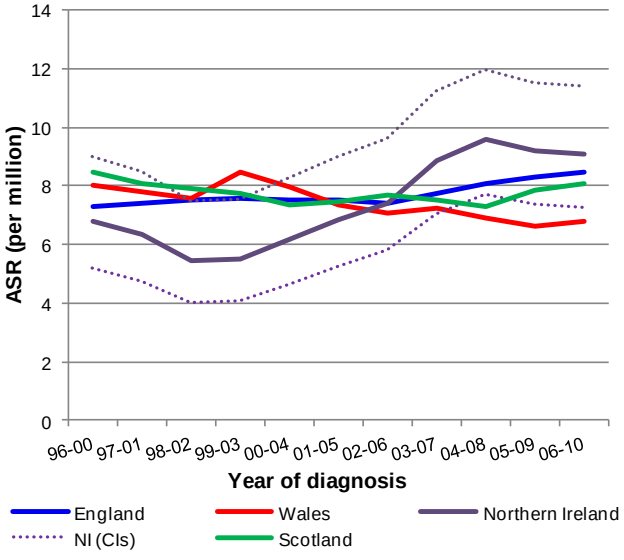
Country	Population	Soft Tissue		Bone	
		No. of tumours	ASR	No. of tumours	ASR
England	52,234,045	2,740	44.9 (43.0 - 47.0)	443	8.2 (8.0 - 9.0)
Scotland	5,222,100	274	43.4 (38.3 - 49.4)	48	8.9 (6.6 - 12.2)
Wales	3,006,430	201	53.4 (46.0 - 62.4)	23	7.1 (4.5 - 11.5)
Northern Ireland	1,799,392	83	46.0 (34.0 - 54.0)	17	9.6 (5.8 - 16.0)
United Kingdom	**62,261,967**	**3,298**	**45.1 (43.5 - 46.7)**	**531**	**8.2 (7.6 - 9.0)**

Figure 3a: Five-year rolling soft tissue sarcoma age-standardised incidence rates for each country (UK: 1996-2010)



CI = 95% confidence intervals

Figure 3b: Five-year rolling bone sarcoma age-standardised incidence rates for each country (UK: 1996-2010)



CI = 95% confidence intervals

Soft tissue sarcoma age-standardised incidence rates did not differ significantly amongst the four UK countries (Figure 3a). Bone sarcoma age-standardised incidence rates did not differ significantly amongst the four UK countries, although oscillations within the Northern Ireland rates could be seen (Figure 3b). This is due to the relatively low number of tumours diagnosed in the much smaller population of Northern Ireland (Table 2).

4.4 Variation in Bone and Soft Tissue Sarcoma Histological Diagnoses with UK Country

Leiomyosarcoma (including GISTs) (22%) and liposarcoma (12%) were the most common specific types of soft tissue sarcoma diagnosed in the UK. A further 20% of sarcomas were diagnosed as "sarcoma; not otherwise specified (NOS)" which could be indicative of either imprecise registration by cancer registries or the inability to make a specific diagnosis. A significantly lower proportion of sarcomas in England were diagnosed as sarcoma NOS compared to the other three UK countries (Figure 4a). There were also differences in the proportion of dermatofibrosarcomas recorded in each of the four UK countries. These tumours accounted for 5% of sarcomas in England, 4% in Wales, 7% in Northern Ireland and 3.5% in Scotland.

Northern Ireland had a higher proportion of rhabdomyosarcomas (RMS), which could be explained by the younger population age structure within Northern Ireland. Northern Ireland also had a significantly lower proportion of myxofibrosarcomas (<1%) compared to the UK average (3%). This could be a reflection of current pathological reporting within Northern Ireland and requires further investigation.

Figure 4a: Proportion of soft tissue sarcoma histological diagnoses in each country (UK: 1996-2010)

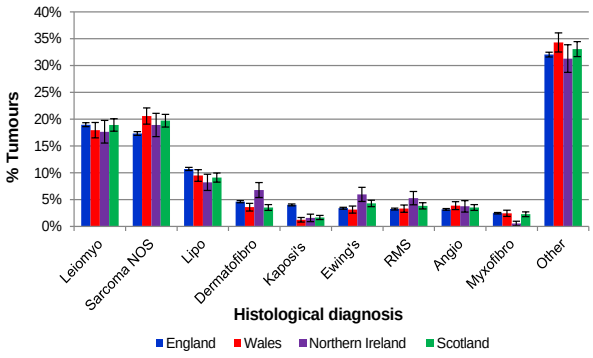
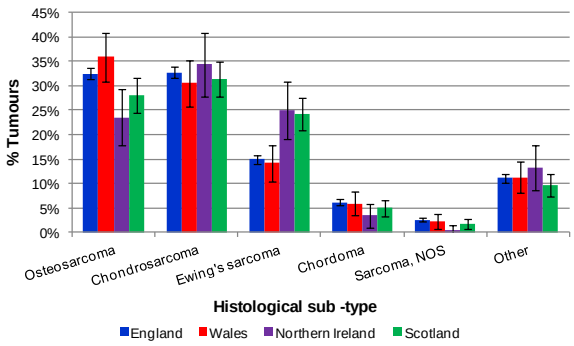


Figure 4b: Proportion of bone sarcoma histological diagnoses in each country (UK: 1996-2010)



Osteosarcoma is frequently cited as the most commonly diagnosed morphological sub-type variant arising in the bones[6]. However, in more recent years, chondrosarcoma appeared to be more commonly diagnosed than osteosarcoma. The proportion of bone sarcomas diagnosed as Ewing's sarcoma was significantly higher in Northern Ireland and Scotland, to the extent that Ewing's sarcomas were more common than osteosarcomas in Northern Ireland. There were no cases of giant cell tumours of the bone recorded in Northern Ireland. This could be a potential data quality issue and requires further investigation (Figure 4b).

4.5 Variation in Bone and Soft Tissue Sarcoma Anatomical Site with UK Country

The most common anatomical cancer sites for soft tissue sarcomas to arise were the extremities (23%), the trunk and thorax (including the intra-abdominal cavity [14%]) and the female genital

tract (11%) (Figure 5a). England had fewer tumours diagnosed within the head and neck region (5%), but a significantly higher proportion of Kaposi's sarcoma (5%). This is due to the high levels of HIV-linked Kaposi's sarcoma diagnosed within the population covered by the London cancer registration office.

Figure 5a: Proportion of soft tissue sarcomas arising at each cancer site in each country (UK: 1996-2010)

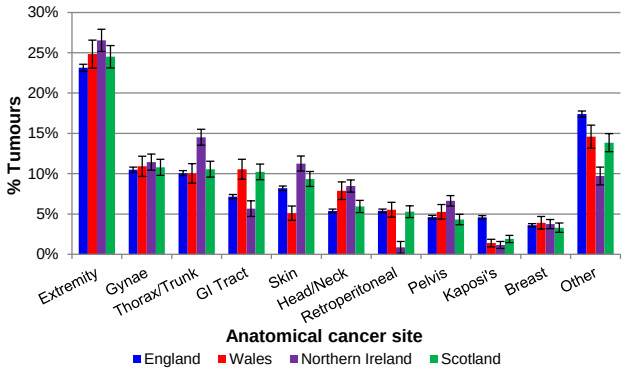
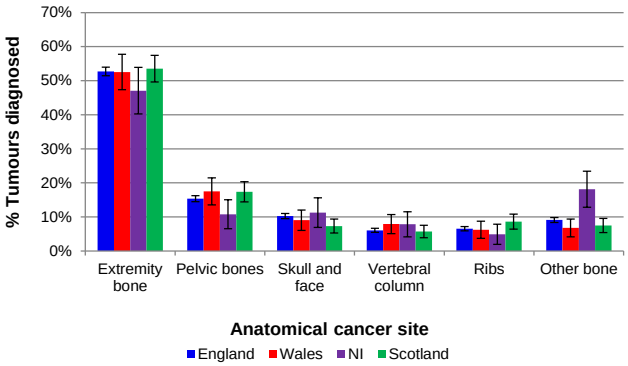


Figure 5b: Proportion of bone sarcomas arising at each cancer site in each country (UK: 1996-2010)



Around 5% of sarcomas arose within the retroperitoneum. This is considerably lower than the 10-20% reported for individual hospital databases. Northern Ireland had a significantly higher proportion of tumours diagnosed within the thorax and trunk (14%), and a significantly lower proportion within the retroperitoneum (<1%). This is due to the tumour coding practice within the Northern Ireland Cancer Registry; retroperitoneal sarcomas are recorded as abdominal tumours (ICD-10 code C494) and consequently included within the thorax and trunk.

The coding of retroperitoneal sarcomas in Northern Ireland is the same as the coding used for retroperitoneal sarcomas by some of the sarcoma specialist centres in England. A national consensus relating to the coding of retroperitoneal sarcomas needs to be agreed if meaningful data on the management of these tumours is to be made available to support centralised specialised commissioning in England. Scotland and Wales had a significantly higher proportion of soft tissue sarcomas arising within the gastro-intestinal tract compared to England and Northern Ireland. Northern Ireland had significantly fewer soft tissue sarcomas recorded as "other" or "unspecified" cancer sites (C498 and C499).

Around 50% of bone sarcomas were diagnosed in the bones of the extremities, 16% within the pelvic bones and 10% within the bones of the skull and face (Figure 5b). There were no significant differences amongst the four UK countries. Northern Ireland had a higher proportion of bone sarcomas within "other" or unspecified bones (18%) and slightly fewer arising within the pelvic bones and extremities. This is a reflection of pathological reporting in Northern Ireland, where more accurate diagnostic information is required.

5.0 SURVIVAL

For the purpose of calculating 5-year relative survival rates, sarcomas arising in the skin were excluded. Skin sarcomas are predominantly dermatofibrosarcomas for which five-year relative survival rates approach 100%.

Overall soft tissue sarcoma 5-year relative survival rates increased significantly between 1996-2000 and 2001-2005 from 50% to 54%. Soft tissue sarcoma 5-year relative survival rates improved from 51% to 55% in males and from 49% to 52% in females (Figure 6a). During the first half of the time period examined there were no significant differences between male and female 5-year relative survival rates for soft tissue sarcoma. However, from the year 2000 onwards, 5-year

relative survival was significantly higher in males than females by a difference of up to four percentage points (Figure 6a). This could result from the increased incidence of well-differentiated liposarcomas in males which have relative survival rates as high as 100% depending on the anatomical cancer site of diagnosis. Bone sarcoma 5-year relative survival rates did not change significantly in the 10-year period studied, and increased by only two percentage points from 54% in 1996-2000 to 56% in 2001-2005. There were no significant differences between the 5-year relative survival rates for bone sarcomas diagnosed in males and females (Figure 6b).

Figure 6a: Soft tissue sarcoma (excluding skin) 5-year rolling 5-year relative survival rates in males and females (UK: 1996-2010)

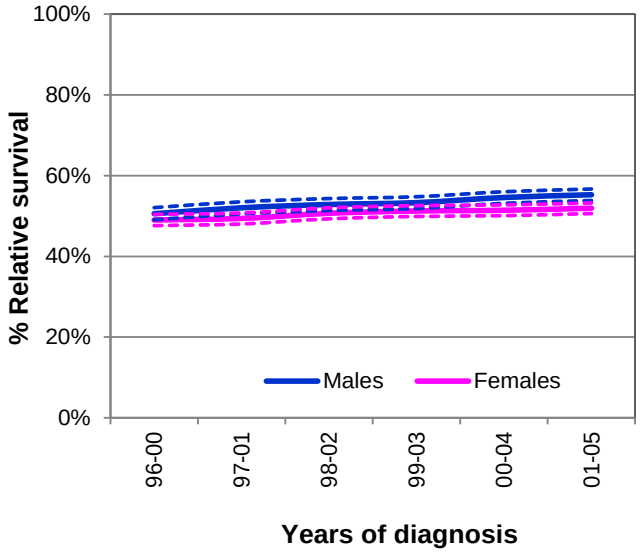


Figure 6b: Bone sarcoma 5-year rolling 5-year relative survival rates in males and females (UK: 1996-2010)

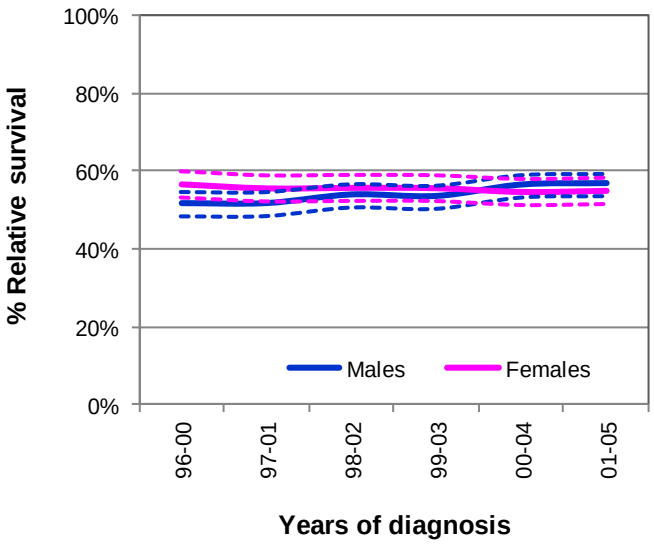


Figure 7a: Soft tissue sarcoma (excluding skin) 5-year rolling 5-year relative survival rates (UK: 1996-2005)

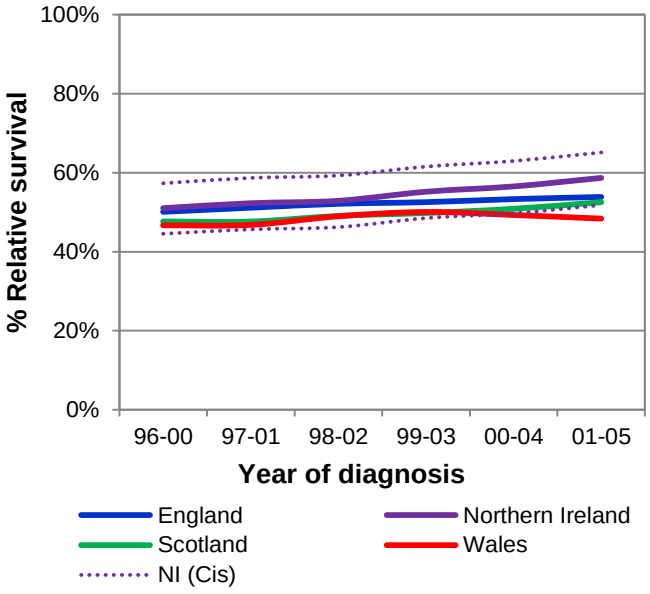
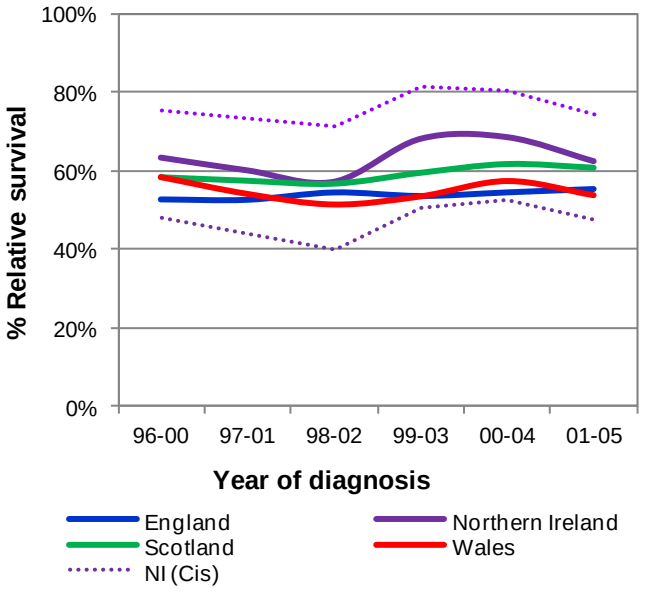


Figure 7b: Bone sarcoma 5-year rolling 5-year relative survival rates (UK: 1996-2005)



CI = 95% confidence intervals

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Throughout the 10-year period studied, soft tissue sarcoma 5-year relative survival rates were always slightly higher in England and Northern Ireland than in Wales and Scotland, but these

differences were not statistically significant (Figure 7a). Bone sarcoma survival rates in Northern Ireland appeared to be higher than those of the other three UK countries (Figure 7b), but this difference was not significant owing to the very small numbers diagnosed.

There are no comparative international studies with which to compare the overall soft tissue sarcoma 5-year relative survival rates found in this study. Survival rates for each histological sub-type and cancer site, are available for bone and soft tissue sarcomas diagnosed in England between 1985 and 2009[7,8]. Bone sarcoma overall 5-year relative survival rates for the UK are considerably lower than the 66% 5-year relative survival rate reported within the "Surveillance Epidemiology and End Results" (SEER) programme (based on 2003-2009)[9].

6.0 CONCLUSIONS

Bone and soft tissue sarcomas are exceptionally rare. This is the first time that incidence and survival statistics for the UK have been published. Soft tissue sarcoma and bone sarcoma age-standardised incidence rates were 45 per million and 8 per million respectively in the period 2006-2010. There were no statistically significant differences in the overall incidence of bone or soft tissue sarcomas or 5-year relative survival rates amongst the four UK countries. There were some differences between UK countries in the types of sarcomas diagnosed and in the underlying anatomical sites. Northern Ireland had significantly higher rates of bone sarcomas arising in other, or unknown, cancer sites. This highlights an area for further investigation to see whether it reflects the need for more detailed pathology reporting or more specific coding by the Northern Ireland Cancer Registry. Northern Ireland had fewer soft tissue sarcomas arising at unspecified cancer sites, and the proportion of myxofibrosarcomas diagnosed in Northern Ireland was significantly lower than that for the other UK countries.
Soft tissue sarcoma 5-year relative survival rates improved significantly between 1996 and 2005, from 51% to 55% for males, and from 49% to 52% for females. Bone sarcoma 5-year relative survival rates remained consistent at around 54%. Overall bone and soft tissue sarcoma 5-year relative survival rates were almost identical amongst the four UK countries. Bone sarcoma survival rates in Northern Ireland appeared higher than those in the three other UK countries, but these differences were not statistically significant owing to the very small numbers diagnosed.

A major limiting factor of the data presented in this report is that the stage of diagnosis for sarcomas diagnosed in the UK was incomplete. It was therefore not possible to adjust the survival analyses to reflect the underlying stage at diagnosis. Staging data will be collected in future in England as part of the new Cancer Outcomes and Services Dataset (COSD) which is now a mandatory requirement for all hospitals treating NHS patients.

REFERENCES

1 World Health Organisation, 2012. International Classification of Diseases (ICD). Available at: http://www.who.int/classifications/icd/en/ [Accessed 24.06.2013].

2 Ederer, F. and Heise, H. 1959. Instructions to Ibm 650 programmers in processing survival computations. Technical, End Results Evaluation Section, National Cancer Institute.

3 Gutierrez, J. C., Perez, E. A., Franceschi, D., Moffat, F. L., Livingstone, A. S. and Koniaris, L. G., 2007. Outcomes for soft-tissue sarcoma in 8,249 cases from a large state cancer registry. Journal of Surgical Research, 141 (1), 105- 114.

4 Nijhuis, P. H. A., Schaapveld, M., Otter, R., Molenaar, W. M., van der Graaf, W. T. A. and Hoekstra, H. J., 1999. Epidemiological aspects of soft tissue sarcomas (STS) - consequences for the design of clinical STS trials. European Journal of Cancer, 35 (12), 1705-1710.

5 Levi, F., La Vecchia, C., Randimbison, L. and Te, V.-C., 1999. Descriptive epidemiology of soft tissue sarcomas in Vaud, Switzerland. European Journal of Cancer, 35 (12), 1711-1716.

6 Dorfman, H. D., Czerniak, B. 1995.Bone Cancers. Interrnational Journal of the American Cancer Society , 75 (Issue Supplement 1), 203-210.

7 Francis et al (2013) Bone sarcoma incidence and survival – Tumours diagnosed between 1985 and 2009 [online], Available: www.ncin.org.uk/view.aspx?rid=1649 [17th April 2013]

8 Francis et al (2013) Soft tissue sarcoma incidence and survival – Tumours diagnosed between 1985 and 2009 [online], Available: www.ncin.org.uk/view.aspx?rid=2062 [17th April 2013]

9 Howlader N, Noone AM, Krapcho M, Garshell J, Neyman N, Altekruse SF, Kosary CL, Yu M, Ruhl J, Tatalovich Z, Cho H, Mariotto A, Lewis DR, Chen HS, Feuer EJ, Cronin KA (eds). SEER Cancer Statistics Review, 1975-2010, National Cancer Institute. Bethesda, MD, http://seer.cancer.gov/csr/1975_2010/, based on November 2012 SEER data submission, posted to the SEER web site, 2013.

APPENDIX A – Comprehensive List of sarcoma Morphology Codes

(WHO 2012. International Classification of Diseases for Oncology, 3rd Edition (ICD-O-3).)

Morphology	Description
8710	Glomangiosarcoma: Glomoid sarcoma
8711	Glomus tumour (nad varients), malignant glomus tumour
8713	Glomangiomyoma
8800	Sarcoma, NOS
8801	Spindle cell sarcoma
8802	Giant cell sarcoma (except of bone M9250/3); pleomorphic cell sarcoma
8803	Small cell sarcoma; round cell sarcoma
8804	Epithelioid sarcoma, epithelioid cell sarcoma
8805	Undifferentiated sarcoma
8806	Desmoplastic small round cell tumour
8810	Fibrosarcoma, NOS, sclerosing epitheliod fibrosarcoma
8811	Fibromyxosarcoma
8812	Periosteal fibrosarcoma (C40._, C41._); periosteal sarcoma, NOS (C40._, C41._)
8813	Fascial fibrosarcoma
8814	Infantile fibrosarcoma; congenital fibrosarcoma
8815	Solitary fibrous tumour, NOS
8821	Aggressive fibromatosis, Desmoid tumour NOS
8822	Abdominal fibromatosis (ICDO-2)
8823	Desmoplastic fibroma (ICD-O-2)
8824	Myofibromatosis (ICD-O3)
8825	Inflammatory myofibroblastic tumour, Myofibroblastic tumour, NOS
8830	Fibrous histiocytoma, malignant; fibroxanthoma, malignant
8832	Dermatofibrosarcoma, NOS (C44._); dermatofibrosarcoma protuberans, NOS (C44._)
8833	Pigmented dermatofibrosarcoma protuberans; Bednar tumour
8834	Giant cell fibroblastoma
8835	Plexiform fibrohistiocytic tumour
8836	Angiomatoid fibrous histiocytoma
8840	Myxosarcoma
8841	Angiomyxoma
8842	Ossifying fibromyxoid tumour, atypical
8850	Liposarcoma, NOS; fibroliposarcoma
8851	Liposarcoma, well differentiated; Liposarcoma, differentiated
8852	Myxoid Liposarcoma; myxoliposarcoma
8853	Round cell liposarcoma
8854	Pleomorphic liposarcoma
8855	Mixed liposarcoma
8857	Fibroblastic liposarcoma
8858	Dedifferentiated liposarcoma
8860	Angiomyoliposarcoma
8890	Leiomyosarcoma, NOS
8891	Epithelioid leiomyosarcoma
8894	Angiomyosarcoma
8895	Myosarcoma
8896	Myxoid leiomyosarcoma
8897	Smooth muscle tumour
8898	Metastasizing leiomyosarcoma
8900	Rhabdomyosarcoma, NOS; rhabdosarcoma
8901	Pleomorphic rhabdomyosarcoma
8902	Mixed type rhabdomyosarcoma

Morphology	Description
8910	Embryonal rhabdomyosarcoma; sarcoma botryoides; botryoid sarcoma
8912	Spindle cell rhabdomyosarcoma
8920	Alveolar rhabdomyosarcoma
8921	Rhabdomyosarcoma with ganglionic differentiation; Ectomesenchymoma
8930	Endometrial stromal sarcoma (C54.1)
8931	Endometrial stromal sarcoma, low grade
8935	Stromal Sarcoma
8936	Gastrointestinal stromal sarcoma
8963	Rhabdoid sarcoma
8964	Clear cell sarcoma of kidney
8982	Myoepithelioma
8990	Mesenchymoma, malignant; mixed mesenchymal sarcoma
8991	Embryonal sarcoma
9020	Phyllodes tumour, malignant (C50.) Cystosarcoma phyllodes, malignant (C50.)
9040	Synovial sarcoma, NOS; synovioma, NOS; synovioma, malignant
9041	Synovial sarcoma, spindle cell
9042	Synovial sarcoma, epithelioid cell
9043	Synovial sarcoma, biphasic
9044	Clear cell sarcoma (except of kidney M8964/3)
9120	Haemangiosarcoma, Angiosarcoma of soft tissue
9130	Haemangioendothelioma, NOS, Kaposiform haemangioepithelioma
9133	Epithelioid haemangioendothelioma, malignant
9135	Endovascular papillary angioendothelioma
9136	Spindle cell hemangioendothelioma
9140	Kaposi sarcoma; Multiple haemorrhagic sarcoma
9150	Haemangiopericytoma, NOS
9170	Lymphangiosarcoma; lymphangioendothelial sarcoma
9174	Lymphangiomyomatosis
9180	Osteosarcoma, NOS (C40._, C41._)
9181	Chondroblastic osteosarcoma (C40._, C41._)
9182	Fibroblastic osteosarcoma (C40._, C41._); osteofibrosarcoma (C40._, C41._)
9183	Telangiectatic osteosarcoma (C40._, C41._)
9184	Osteosarcoma in Paget's disease of bone (C40._, C41._)
9185	Small cell osteosarcoma (C40._, C41._)
9186	Central osteosarcoma (C40._, C41._);
9187	Intraosseous well differentiated osteosarcoma (C40._, C41._)
9190	juxtacortical osteosarcoma ICD-O-2
9192	Parosteal osteosarcoma (C40._, C41._)
9193	Periosteal osteogenic sarcoma (C40._, C41._)
9194	High grade surface osteosarcoma (C40._, C41._)
9195	Intracortical osteosarcoma (C40._, C41._)
9200	Aggressive osteoblastoma
9210	Osteochondromatosis
9220	Chondrosarcoma
9221	Juxtacortical chondrosarcoma (C40._, C41._)
9230	Chondroblastoma, malignant (C40._, C41._)
9231	Myxoid chondrosarcoma
9240	Mesenchymal chondrosarcoma
9242	Clear cell chondrosarcoma, (C40._, C41._)
9243	Dedifferentiated chondrosarcoma (C40._, C41._)
9250	Giant cell tumour of bone, NOS
9251	Giant cell tumour of soft parts, NOS

Morphology	Description
9252	Malignant tenosynovial giant cell tumour (C49._)
9260	Ewing's sarcoma, Ewing's tumour, Extraskeletal Ewing tumour
9261	Adamantinoma of long bones; tibial adamantinoma (C40.2)
9270	Odontogenic tumour
9290	Ameloblastic odontosarcoma: Ameloblastic fibrodentinosarcoma
9310	Ameloblastoma
9330	Ameloblastic fibrosarcoma: Ameloblastic sarcoma: Odontogenic fibrosarcoma
9341	Clear cell odontogenic tumour
9342	Odontogenic carcinosarcoma
9364	Peripheral neuroectodermal tumour; neuroectodermal tumour, NOS
9365	Askin tumour
9370	Chordoma
9371	Chondroid chordoma
9372	Dedifferentiated chordoma
9373	Parachondroma
9473	Primitive neuroectodermal tumour
9540	Malignant peripheral nerve sheath tumour MPNST, NOS
9560	Malignant schwannoma; neurilemoma, malignant
9561	Malignant peripheral nerve sheath tumour with rhabdomyoblastic differentiation
9571	Perineurioma, malignant; Perineural MPNST
9580	Granular cell tumour, malignant; granular cell myoblastoma, malignant
9581	Alveolar soft part sarcoma

APPENDIX B – ICD-10 Site Codes Included Within Each Anatomical Cancer Site

Site	ICD-10 codes
Limb	C491 (connective and soft tissue of upper limb), C492 (connective and soft tissue of lower limb), C471 (peripheral nerves of upper limb), C472 (peripheral nerves of lower limb), C764 (other and ill defined sites: upper limb), C765 (other and ill defined sites: lower limb)
Head, face and neck	C00 (lip), C01 (base of tongue), C02 (other and unspecified parts of tongue), C03 (gum), C04 (floor of mouth), C05 (palate), C06 (other and unspecified parts of mouth), C07 (parotid gland), C08 (other and unspecified major salivary glands), C09 (tonsil), C10 (oropharynx), C11 (nasopharynx), C12 (pyriform sinus), C13 (hypopharynx), C14 (other and ill-defined sites in the lip oral cavity and pharynx), C30 (nasal cavity and middle ear), C31 (accessory sinuses), C32 (larynx), C69 (eye and adnexa), C490 (connective and soft tissue of head, face and neck), C760 (other and ill defined sites: head, face and neck), C761 (other and ill defined sites: thorax)
Retroperitoneum	C48 (retroperitoneum and peritoneum)
Connective tissue of trunk	C493 (connective and soft tissue of thorax), C494 (connective and soft tissue of abdomen), C495 (connective and soft tissue of pelvis), C496 (connective and soft tissue of trunk), C762 (other and ill defined sites: abdomen), C763 (other and ill defined sites: pelvis)
GI tract	C15 (oesophagus), C16 (stomach), C17 (small intestine), C18 (colon), C19 (rectosigmoid junction), C20 (rectum), C21 (anus and anal canal)
Organs within the trunk	C22 (liver and intrahepatic bile ducts), C23 (gallbladder), C24 (other and unspecified parts of biliary tract), C25 (pancreas), C26 (other and ill-defined digestive organs), C33 (trachea), C34 (bronchus and lung), C37 (thymus), C38 (heart, mediastinum and pleura), C39 (other and ill-defined sites in the respiratory system and intrathoracic organs), C64 (kidney, except renal pelvis), C65 (renal pelvis), C66 (ureter), C67 (bladder), C68 (other and unspecified urinary organs), C73 (thyroid gland), C74 (adrenal gland), C75 (other endocrine glands and related structures)
Skin	C44 (other malignant neoplasms of skin, not malignant melanoma)
Kaposi's sarcoma	C46 (Kaposi's sarcoma)
Peripheral nerves	C470 (peripheral nerves of head, face and neck), C473 (peripheral nerves of thorax), C474 (peripheral nerves of abdomen), C475 (peripheral nerves of pelvis), C476 (peripheral nerves of trunk, unspecified), C478 (overlapping lesion of peripheral nerves and autonomic nervous system), C479 (peripheral nerves and autonomic nervous system, unspecified)
Brain	C70 (meninges), C71 (brain), C72 (spinal cord, cranial nerves and other parts of the central nervous system)
Breast	C50 (breast)
Genital organs	C51 (vulva), C52 (vagina), C53 (cervix uteri), C54 (corpus uteri), C55 (uterus, part unspecified), C56 (ovary), C57 (other and unspecified female genital organs), C58 (placenta), C60 (penis), C61 (prostate), C62 (testis), C63 (other and unspecified male genital organs)
Gynaecological organs	C51 (vulva), C52 (vagina), C53 (cervix uteri), C54 (corpus uteri), C55 (uterus, part unspecified), C56 (ovary), C57 (other and unspecified female genital organs), C58 (placenta)
Male genitalia	C60 (penis), C61 (prostate), C62 (testis), C63 (other and unspecified male genital organs)
Other/Unknown	All others