Bone Sarcoma Incidence and Survival

Tumours Diagnosed Between 1985 and 2009

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EXECUTIVE SUMMARY

Primary bone sarcomas are an exceptionally rare form of cancer, accounting for only 0.2% of all malignancies diagnosed in England. This report is designed to fill a void in publicly available data describing the incidence and survival of patients diagnosed with primary bone sarcoma. The report considers the four most common variants of bone sarcoma and presents incidence and survival data according to the age of the patient at diagnosis and the cancer site for all sarcomas and for the four variants. Survival rates are also presented for the main anatomical sub-sites.

The average annual incidence of bone sarcoma in England over the period 1985-2009 is around 7.9 per million persons. This has increased in more recent years to around 10 per million persons, but this may reflect improved diagnostic techniques and reporting rather than a true increase in incidence. There were 437 new diagnoses of bone sarcoma in 2009; 131 (30%) were osteosarcomas, 161 (37%) were chondrosarcomas, 28 (6%) were chordomas and 62 (14%) were Ewing's sarcomas.

Ewing's sarcoma and osteosarcoma are the most common forms of bone sarcoma diagnosed in children, teenagers and young adults. Chordomas and chondrosarcomas are more common in the elderly but are exceptionally rare in children and young adults. The incidence of osteosarcoma is bimodal, making it the only variant relatively common in both teenagers and the elderly. Chondrosarcomas are the most common form of bone sarcoma diagnosed in patients over the age of 35 years.

The four major variants of bone sarcoma are more common in males than in females. Over 90% of bone sarcomas diagnosed in patients under the age of 19 are either osteosarcomas or Ewing's sarcomas. From the age of 20 years and onwards, chondrosarcomas become more commonly diagnosed, and this variant accounts for fifty percent of tumours diagnosed in patients aged 50 to 59 years. Rarer tumours, such as malignant fibrous histiocytoma and ameloblastoma, are most likely to arise in patients aged 40 years and above.

The most common sites for bone sarcoma to arise are within the bones of the lower limbs (38%), the pelvic bones (16%) and the bones of the upper limbs (14%). As patients become older, a smaller proportion of tumours arise in the limbs, and tumours of the pelvis become more common. Around 70% of bone sarcomas diagnosed in patients under the age of 20 years arise in the limbs, compared to around 40% in patients aged 40 years and over. The proportion of bone sarcomas diagnosed in the skull and face is greatest in patients aged between 30 and 59 years, and the proportion where the site is not specified increases with age

There have been no significant improvements in bone sarcoma 5-year relative survival rates for patients diagnosed between 1985 and 2004. At around 54%, relative survival rates in England are low when compared with those calculated using other international databases such as the Surveillance, Epidemiology and End Results" (SEER) programme. Differences in the age distribution of patients included in the two cohorts and the variants included in the analyses could explain some of these differences.

Further survival analyses are required to consider the stage of the tumour at diagnosis, as well as how the stage varies with age and deprivation for the different types of bone sarcoma. Further analyses are also required to investigate the incidence of osteosarcoma bone tumours diagnosed in the elderly, and to establish whether these tumours are radiation induced or related to a history of Paget's disease.

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1.0 INTRODUCTION

Primary bone sarcomas are an exceptionally rare form of cancer, accounting for only 0.2% of all malignancies diagnosed in England. Primary bone cancers represent a challenge as they are rare and best practice guidelines state that they should be treated within high caseload centres. There are many different types of bone sarcoma, of which osteosarcoma, chondrosarcoma, chordoma and Ewing's sarcoma are the most common and collectively account for over 85% of all tumours diagnosed. Other relatively uncommon bone sarcomas account for approximately 6% of malignancies diagnosed in patients under the age of twenty. There are few reports which examine the incidence and outcome of patients with a primary bone cancer. This report considers the four most common variants of bone sarcoma and presents incidence and survival data according to the age of the patient at diagnosis and the cancer site. Survival rates are also presented for the main anatomical sub-sites.

2.0 METHODS AND DATA

The West Midlands Cancer Intelligence Unit (WMCIU) is the National Cancer Intelligence Network (NCIN) lead registry in England for bone and soft tissue sarcoma. The lead registry 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 regional cancer registries which covers all cases diagnosed in England. The current version of the NCDR includes all malignancies diagnosed in England between 1985 and 2009. There are potential coding issues with regard to the "older" tumours within the NCDR, but utilising the whole dataset allows observation of temporal changes in the incidence of bone sarcomas to be made.

Bone sarcomas were classified using the ICD-10 coding system – all tumours where the first three digits of the site codes are C40 (Malignant neoplasm of bone and articular cartilage of limbs) or C41 (Malignant neoplasm of bone and articular cartilage of other and unspecified sites) are included in this report. In most cases, incidence data are reported as three-year rolling averages to smooth the apparent oscillations caused by the relatively small number of bone sarcomas involved. Confidence intervals around the incidence rates were calculated using the gamma method[1]. 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. 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

When malignant tumours arise in the bones, the tumours are primary tumours which are classified as bone sarcomas. When cancer cells spread to the bones from cancers in other sites in the body, they can form secondary bone metastases. There are data quality issues within the NCDR, as tumours exist which are coded as either C40 or C41 with a carcinoma rather than a sarcoma morphology code. These tumours are most probably not primary bone cancers, but secondary bone metastases where the underlying cancer site is unknown. These data quality issues are discussed in more detail in the bone sarcoma data quality report[2]. For the purpose of the analyses within this report, only tumours coded as either C40 or C41 and with a sarcoma morphology code are included.

[1] Confidence Intervals for directly standardised rates: a method based on the Gamma Distribution, Stat Med, 1997, 791-801

[2] http://www.ncin.org.uk/search/bone+sarcoma+completeness.aspx

Between 1985 and 2009, 10,605 tumours were registered as bone sarcomas, of which 9,573 (90%) were registered with a sarcoma morphology code, 332 with a carcinoma morphology code and 700 (7%) as "Neoplasm, malignant", where the true morphology of the tumour is unknown. Tumours with carcinoma and unknown morphology type were removed from the dataset, leaving 9,573 bone sarcomas available for analysis.

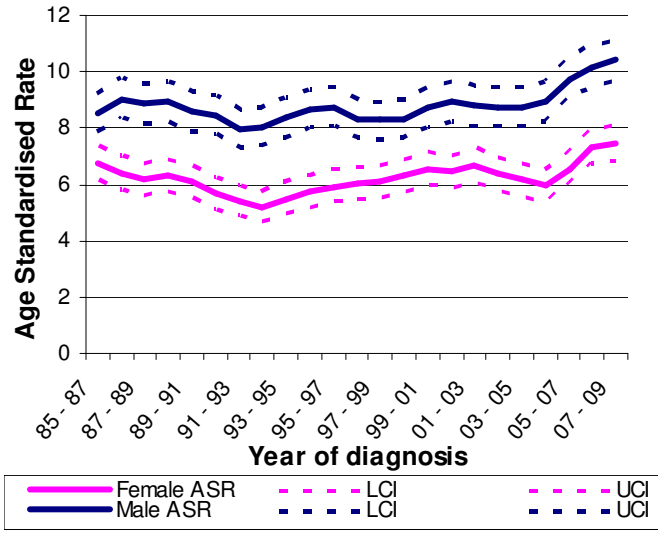
4.0 RESULTS

4.1 All Malignant Primary Bone Cancers

4.1.1 Incidence

9,573 primary bone sarcomas were diagnosed in England between 1985 and 2009. The average annual incidence rate over the period 1985-2009 is around 7.9 per million persons, increasing to approximately 10 per million in more recent years. There were 486 new diagnoses of bone sarcoma in 2008 and 437 in 2009. Age standardised incidence rates are significantly higher in males than in females (Figure 4.1.1). Although it appears that the annual incidence of bone cancer may be increasing, this is probably a reflection of improved diagnostic techniques and reporting.

Figure 4.1.1: Bone sarcoma 3-year rolling age standardised incidence rates (England: 1985 – 2009)



4.1.2 Variation in Incidence with Morphological Sub-type

Figure 4.1.2a: Age specific rates of most common bone sarcoma variants (England: 1985 – 2009)

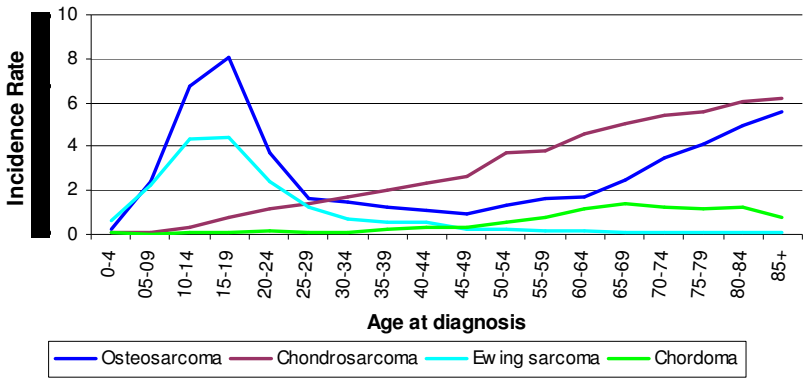
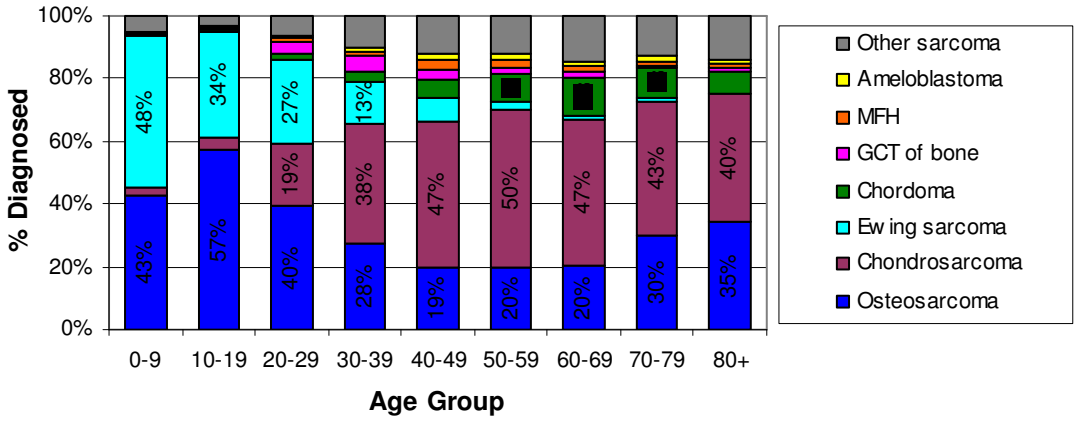


Figure 4.1.2a shows how the incidence of the four most common sub-types of bone sarcoma vary across age groups. Ewing's sarcoma and osteosarcoma are the most common forms of bone sarcoma diagnosed in children, teenagers and young adults. In contrast, chordomas and chondrosarcomas are more common in the elderly but are exceptionally rare in children and young adults. The incidence of osteosarcoma is bimodal, making osteosarcoma the only variant relatively common in both teenagers and the elderly. Chondrosarcomas are the most common form of bone sarcoma diagnosed in patients over the age of 35 years.

Although the four most common variants of bone sarcoma account for over 85% of all diagnoses, other types can arise, such as giant cell tumour (GCT) of the bone, malignant fibrous histiocytoma (MFH), and ameloblastoma. Figure 4.1.2b shows how the types of bone sarcoma diagnosed vary with age. Over 90% of bone sarcomas diagnosed in patients under the age of 19 years are either osteosarcomas or Ewing's sarcomas. From the age of 20 years and onwards, chondrosarcomas become more commonly diagnosed, and this variant accounts for 50% of tumours diagnosed in patients aged 50 to 59 years. Rarer tumours, such as MFH and ameloblastoma, are most likely to arise in patients aged 40 years and above.

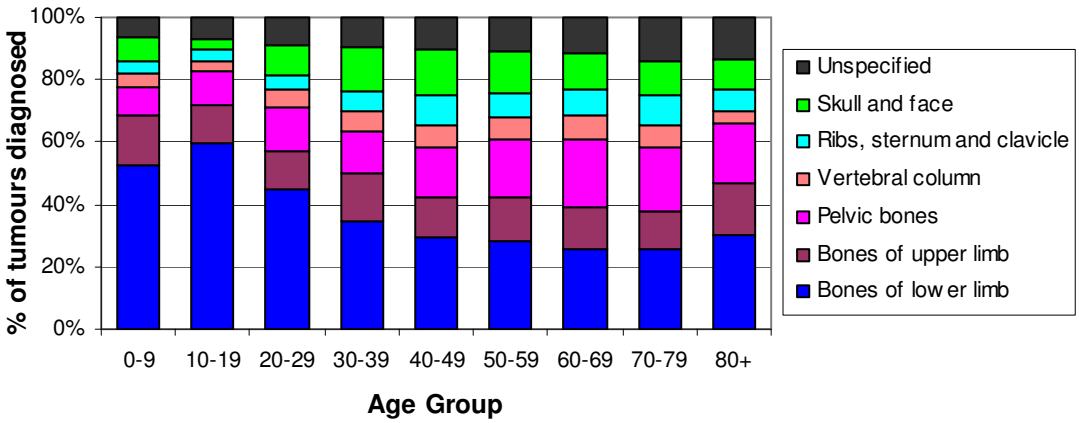
Figure 4.1.2b: Proportion of bone sarcoma variants diagnosed by age group and morphology (England: 1985 – 2009)



4.1.3 Variation in incidence with anatomical site

Primary bone cancer can arise within any bone of the human body and can affect patients of any age. The most common sites for bone sarcoma to arise are within the bones of the lower limbs (38%), the pelvic bones (16%) and the bones of the upper limbs (14%). As patients become older, a smaller proportion of tumours arise in the limbs, and tumours of the pelvis become more common.

Figure 4.1.3: Proportion of bone sarcomas diagnosed by age group and anatomical site (England: 1985 – 2009)

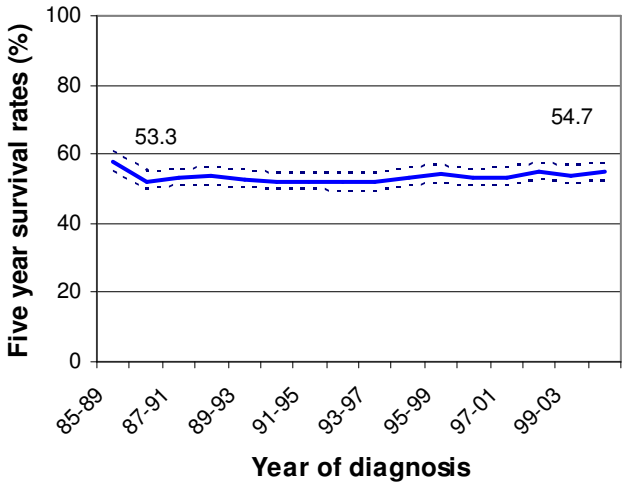


The site of diagnosis is dependent on age, with around 70% of bone sarcomas diagnosed in patients under the age of 20 years arising in the limbs, compared to around 40% for patients aged 40 years and over (Figure 4.1.3). The proportion of bone sarcomas diagnosed in the skull and face is greatest in patients aged between 30 and 59 years, and the proportion of bone sarcomas where the site is not specified increases with age.

4.1.4 Survival

There have been no significant improvements in overall 5-year relative survival rates for patients with bone sarcoma over the past twenty five years, with rates oscillating between 53% and 55% (Figure 4.1.4). The overall 5-year relative survival for patients diagnosed in England in 1985-2004 are considerably lower than the 66% 5-year relative survival rates reported within the "Surveillance, Epidemiology and End Results" (SEER) programme[3] (based on 2002-2008 data, where 1975 to 2009 data is available). The age distribution of patients could explain some of this difference. Thirty percent of patients diagnosed with a bone sarcoma in England were over the age of 65, compared with 21% within the SEER data, and 22% were under the age of 19 compared with 29% in the SEER data. Also, the SEER data do not specify which morphological sub-types are included within the analyses. Further analyses are thus required before any conclusions can be made for the differing survival rates.

Figure 4.1.4: 5-year bone sarcoma relative survival rates (England: 1985 – 2004)



4.2 Osteosarcoma

Osteosarcomas are the most common bone sarcomas accounting for approximately 30% of all diagnoses, and are renowned for being highly malignant. Twelve different variants of osteosarcoma were recorded in the NCDR between 1985 and 2009. The most commonly recorded histological sub-type was M9180 (osteosarcoma, NOS) which accounted for 85% of all osteosarcoma diagnoses. The next most common type was M9181 (chondroblastic osteosarcoma) of which there were only 171 diagnoses between 1985 and 2009. For the purposes of the following analyses, the incidences of all osteosarcoma sub-types have been aggregated.

4.2.1 Osteosarcoma incidence

Between 1985 and 2009, 3,283 osteosarcomas of the bone were diagnosed with an average of 135 tumours diagnosed each year between 2000 and 2009. The age standardised osteosarcoma incidence rate fluctuates around 2.5 per million (Figure 4.2.1a). Osteosarcoma incidence rates are

[3] http://seer.cancer.gov/statfacts/html/bones.html

significantly higher in males than in females with age standardised rates of approximately 3.5 per million in males and 2.5 per million in females in 2007-2009 (Figure 4.2.1b).

Figure 4.2.1a: 3-year rolling age standardised osteosarcoma incidence rates (England: 1985 – 2009)

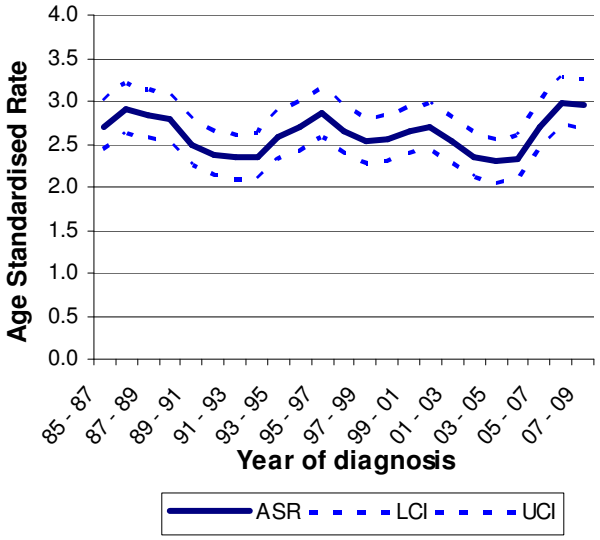
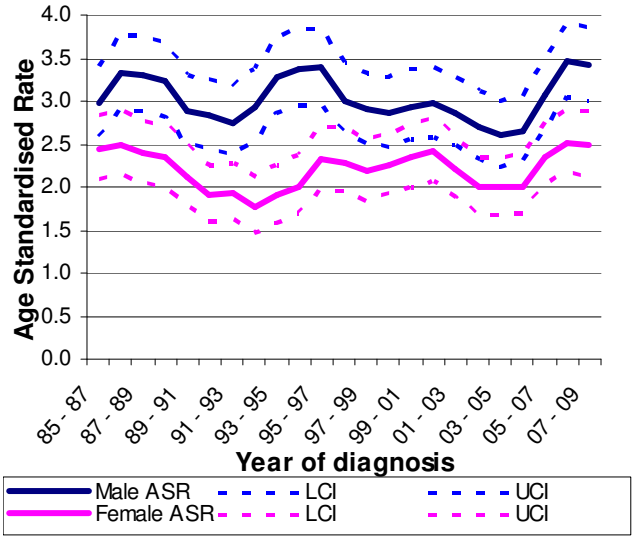
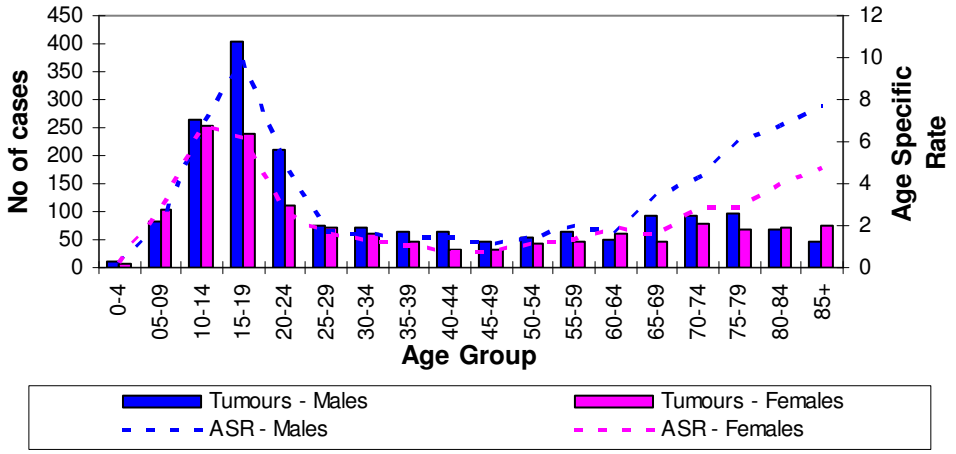


Figure 4.2.1b: 3-year rolling age standardised osteosarcoma incidence rates (England: 1985 – 2009)



Age specific osteosarcoma incidence rates are generally bi-modal, with the first peak appearing before 20 years of age, and a second peak in the elderly (Figure 4.2.1c). Age specific rates are 1.7 times higher in males aged 15 to 24 years than in females in the same age group. Age specific rates are highest in males aged 15-19 years (9.9 per million) and in males aged 85 years and over (7.8 per million). Further analyses are required to investigate the incidence of osteosarcomas diagnosed in the elderly, and to establish whether these tumours are radiation induced or related to a history of Paget's disease

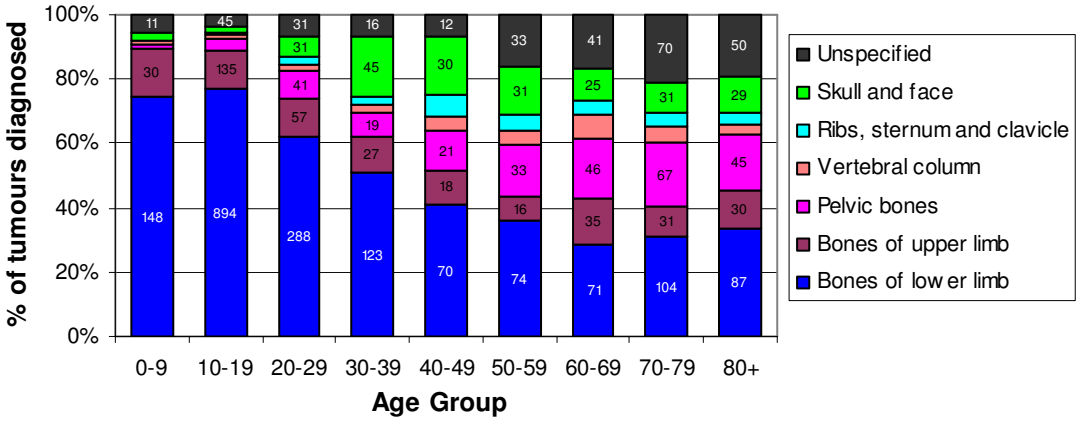
Figure 4.2.1c: Age specific osteosarcoma rates (England: 1985 – 2009)



4.2.2 Variation in osteosarcoma incidence with anatomical site

Figure 4.2.2 shows the proportion of osteosarcomas diagnosed in each age group and at each anatomical site. Osteosarcomas are most predominant in the limbs for patients under the age of 40 years, after which they become more commonly diagnosed in the pelvic bones. Potential coding issues also arise in elderly patients, with over 20% of osteosarcoma patients aged 70 years and above diagnosed with osteosarcoma at an unspecified site. Patients aged between 30 and 60 years are most predisposed to a diagnosis of osteosarcoma of the bones of skull or facial skeleton.

Figure 4.2.2: Proportion of osteosarcomas diagnosed by age group and anatomical site (England: 1985 – 2009)



4.2.3 Osteosarcoma survival

Five-year relative survival for patients with an osteosarcoma has remained static over the past 25 years and is still only around 42% (Figure 4.2.3).

Figure 4.2.3: 5-year osteosarcoma relative survival rates by anatomical site (England: 1985 – 2004)

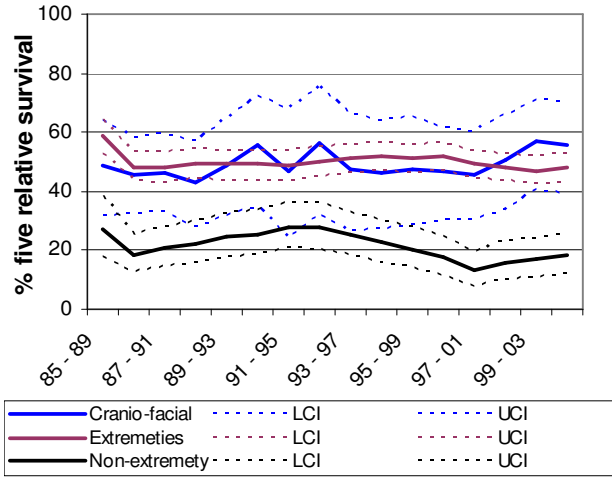


Figure 4.2.4a: 5-year osteosarcoma relative survival rates by age at diagnosis (England: 1985 – 2004)

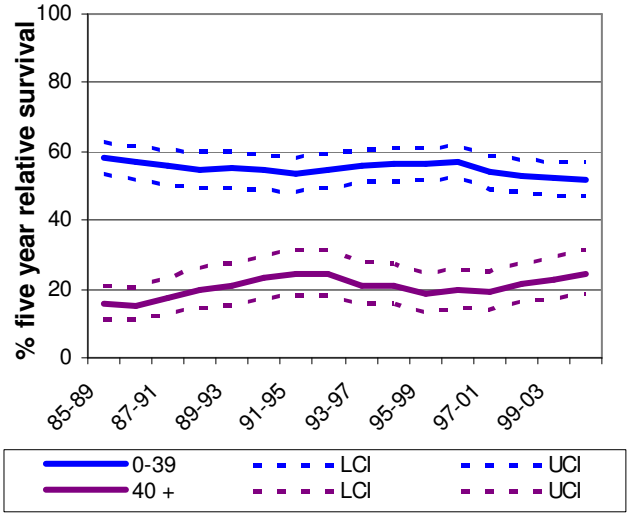
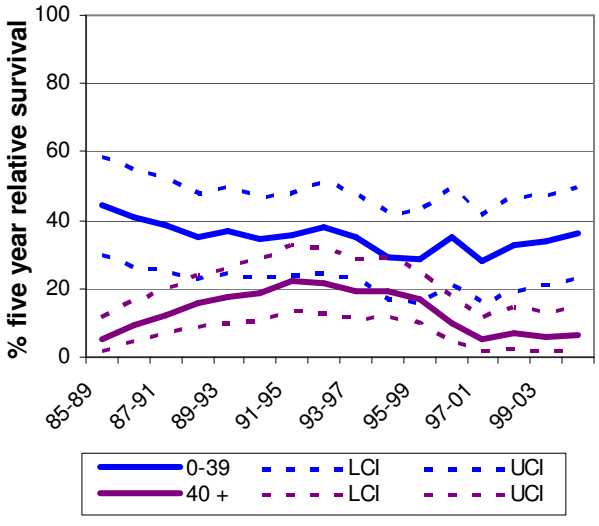


Figure 4.2.4b: 5-year non-extremity osteosarcoma relative survival rates (England: 1985 – 2004)



Five-year relative survival rates are significantly higher in patients under the age of 40 years (52%) when compared with patients over the age of 40 years (25%) (Figure 4.2.4a). Relative survival rates for patients with an osteosarcoma of the non-extremities are relatively poor, with patients under the age of 40 years having a relative survival of 36% and those aged over 40 years having a relative survival as low as 6%.

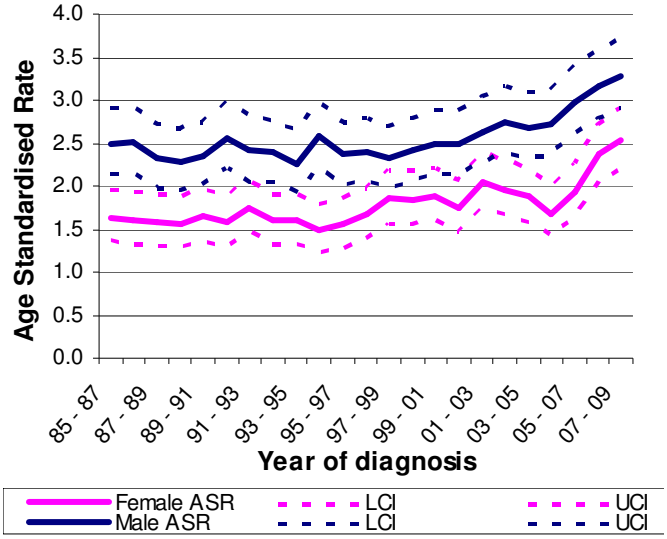
4.3 Chondrosarcoma

Chondrosarcomas are tumours which arise in the cartilage. Chondrosarcoma is the second most common form of bone sarcoma. Eight different variants of chondrosarcoma were recorded in the NCDR between 1985 and 2009. The most common histological sub-type was M9220 (Chondrosarcoma, NOS) which accounted for 91%, followed by M9231 (Myxoid chondrosarcoma) which accounted for 163 (5%) of the 2,954 chondrosarcoma diagnoses. For the purposes of the following analyses, the incidences of all chondrosarcoma sub-types were aggregated.

4.3.1 Chondrosarcoma incidence

Between 1985 and 2009 there were 2,954 diagnoses of chondrosarcoma and these accounted for 31% of all bone sarcomas. Approximately 145 new cases of chondrosarcoma are diagnosed annually. In 2009, there were 161 chondrosarcomas diagnosed in England, resulting in an age standardised incidence rate of approximately 2.9 per million persons. Incidence rates are significantly higher in males than in females with an age standardised rate of approximately 3.3 per million in males and 2.5 per million in females in 2007-2009 (Figure 4.3.1a).

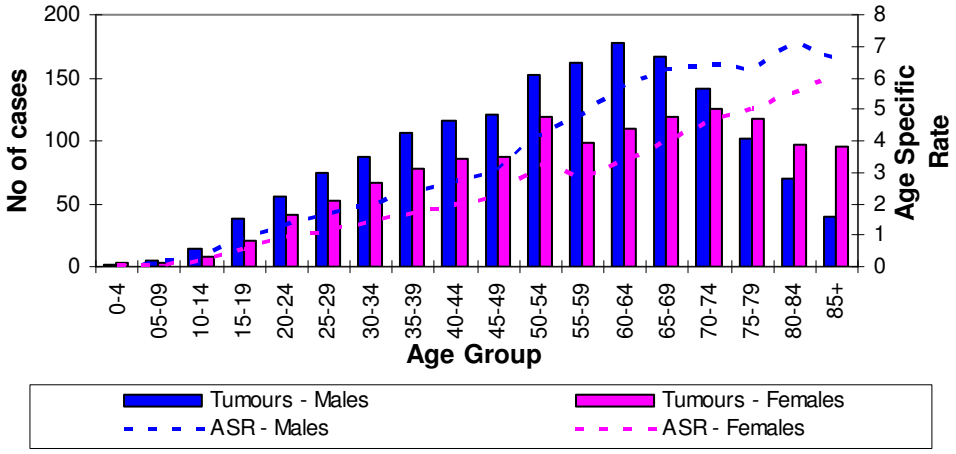
Figure 4.3.1a: 3-year rolling age standardised chondrosarcoma incidence rates by sex England: 1985 – 2009)



Chondrosarcoma incidence rates appear to have increased significantly between 2000-2002 and 2007-2009, but this could either be a reflection of improved diagnostic techniques and reporting or a change in criteria for diagnosing these tumours.

The incidence rates of chondrosarcoma increase with age, and these tumours are exceptionally rare in patients under the age of 15 years. Age specific rates are considerably higher in males than females, with a ratio of 1.5:1.0 in males and females aged between 55 and 84 years (Figure 4.3.1b). Rates are at the highest in males aged 80 years and over, where they are approximately 7 per million.

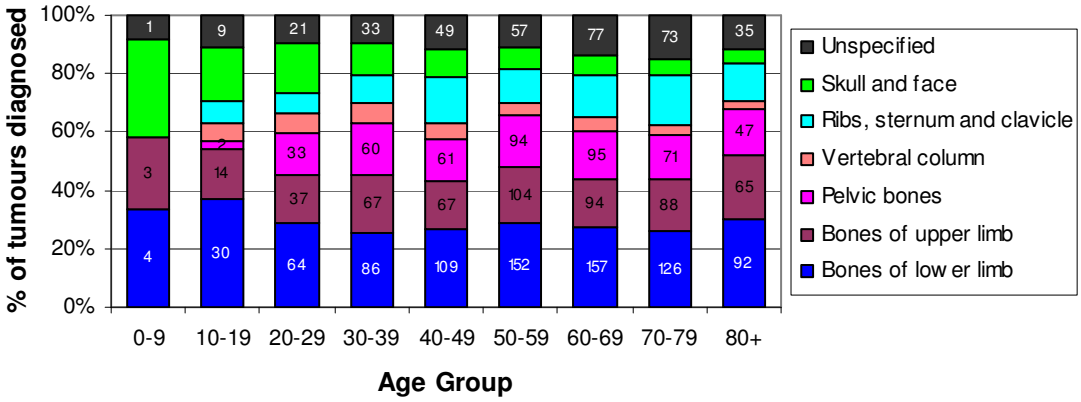
Figure 4.3.1b: Age specific chondrosarcoma rates by sex (England: 1985 – 2009)



4.3.2 Variation in chondrosarcoma incidence with anatomical site

Figure 4.3.2 shows the proportion of chondrosarcomas diagnosed in each age group and at each anatomical site. Chondrosarcomas are most likely to arise within the extremities and pelvic bones, where they account for 62% of all diagnoses. Interestingly, the anatomical site of diagnosis does not generally appear to vary with age. Although Figure 4.3.2 appears to suggest that children are more predisposed to a diagnosis of chondrosarcoma of the skull, only twelve children under the age of ten were diagnosed with a chondrosarcoma between 1985 and 2009. Unlike osteosarcoma diagnoses, the proportion of chondrosarcomas recorded with an unspecified site of diagnosis does not vary with age.

Figure 4.3.2: Proportion of chondrosarcomas diagnosed by age and anatomical sub-site (England: 1985 – 2009)



4.3.3 Chondrosarcoma survival

Five-year relative survival rates are significantly higher for patients with a chondrosarcoma (68%) than for osteosarcoma patients (43%). Five-year relative survival for chondrosarcomas has not improved over the last twenty five years (Figure 4.3.3a). Patients with cranio-facial or extremity chondrosarcoma have better relative survival rates than patients with non-extremity tumours (Figure 4.3.3b). Five-year relative survival rates for patients with non-extremity chondrosarcomas fluctuate around 60%, which is significantly higher than non-extremity osteosarcoma survival rates (approximately 18%).

Figure 4.3.3a: 5-year chondrosarcoma relative survival rates (England: 1985 – 2004)

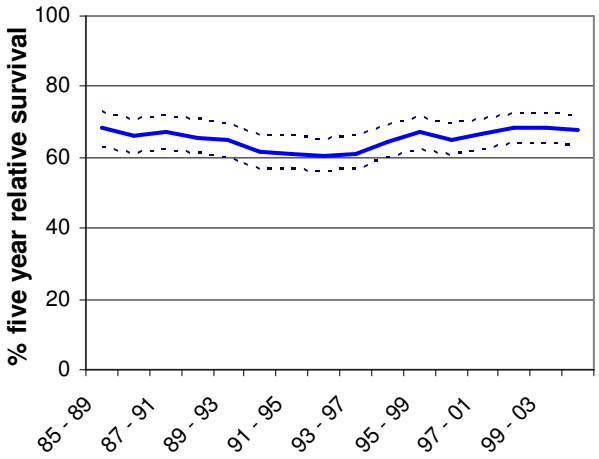
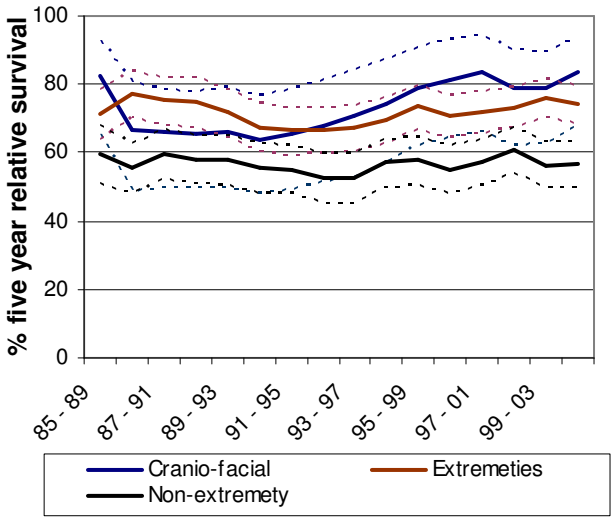
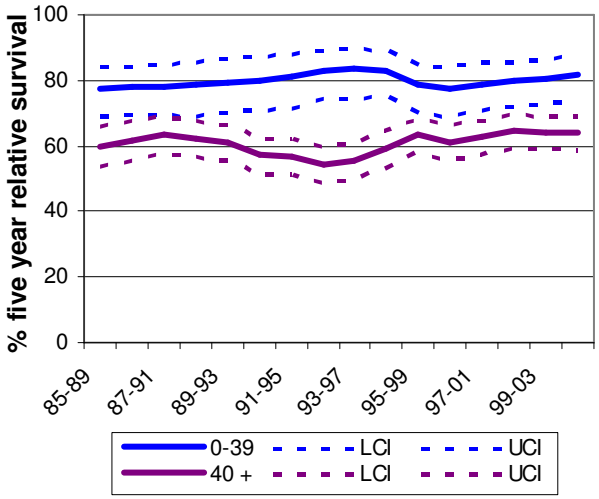


Figure 4.2.4a: 5-year chondrosarcoma relative survival rates by site of diagnosis (England: 1985 – 2004)



Patients with chondrosarcomas of the cranio-facial skeleton also appear to have better relative survival rates than patients with non-extremity chondrosarcomas. However, these analyses are based on exceptionally rare tumours and there is no significant difference in cranio-facial and non-extremity chondrosarcoma relative survival rates.

Figure 4.3.4: Five year chondrosarcoma survival rates by age at diagnosis (England: 1985 – 2004)



Five-year chondrosarcoma relative survival rates are significantly higher in patients diagnosed under the age of 40 years, although the differences in age group survival are not as great as with osteosarcomas. Chondrosarcoma relative survival rates are in excess of 60% for patients in each age group.

4.4 Chordoma

Chordomas are exceptionally rare bone tumours arising within the notochord. There are no population based incidence studies available for chordomas, other than those published based on the SEER program database and old Finish data (1958 to 1971)[4]. There are also a few cohort

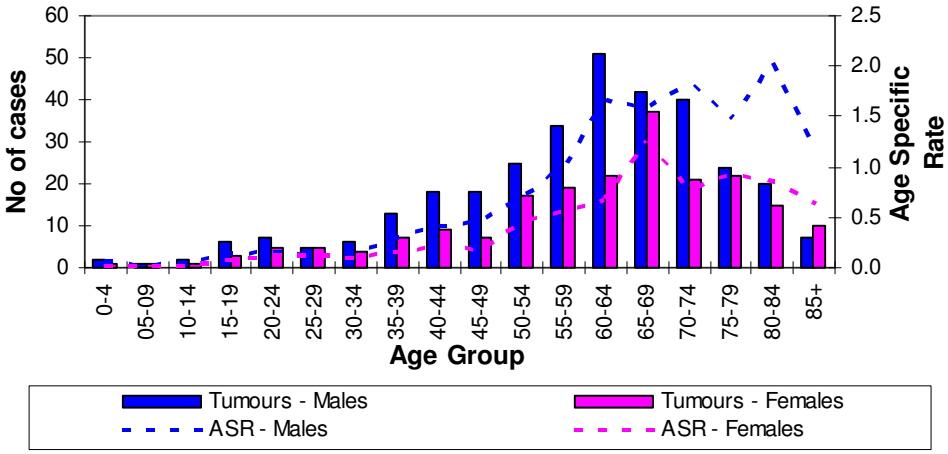
[4] Chordoma in Finland, Acta Orthopaedica, Informa Healthcare

based reports (such as skull base[5]). Therefore, there appears to be a gap in the current literature regarding the annual incidence and survival of patients diagnosed with these rare tumours.

4.4.1 Chordoma incidence

Approximately 25 chordomas are diagnosed annually in England and the age standardised incidence rate is 0.5 per million persons. Chordomas are rarely diagnosed in children and are far more predominant in males over the age of 60 years. In 1985-2009 over 77% of chordomas were diagnosed in patients aged 50 years and over.

Figure 4.4.1a: Age specific chordoma incidence rates (England: 1985 – 2009)



The age-adjusted incidence rate for chordomas of 0.45 per million in England in 1985-2099 is lower than in the 0.8 per million United States data (1973-1995) reported by McMaster et al (2001)[6]. This study also reported the incidence of chordomas to be age dependent, and much more common in males.

4.4.2 Variation in chordoma incidence with anatomical site

Figure 4.4.1a: Proportion of tumours diagnosed by age and anatomical sub-site (England: 1985 – 2009)

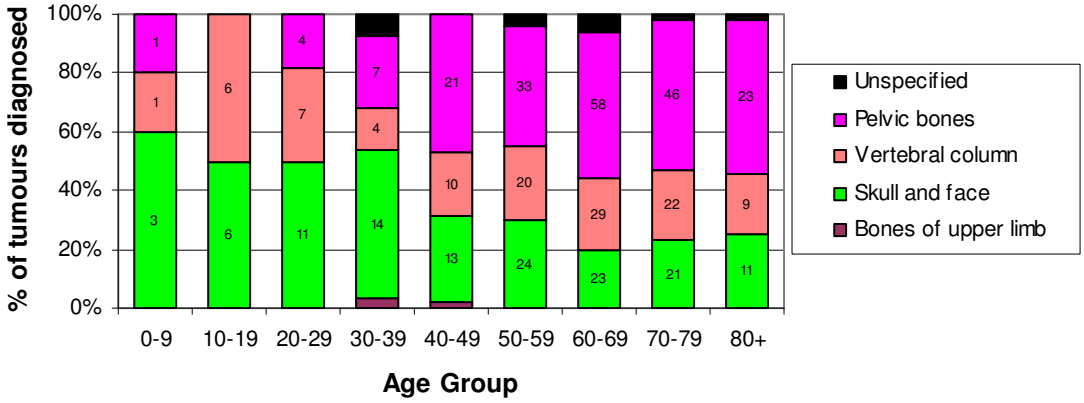


Figure 4.4.1a shows the proportion of chordomas diagnosed in each age group and at each anatomical site. Chordoma diagnoses are almost entirely confined to the pelvic bones, vertebral

[5] Base of skull chordoma. A correlative study of histologic and clinical features of 62 cases - O'Connell - 2006 - Cancer - Wiley Online Library

[6] Chordoma: incidence and survival patterns in the United States (1973 to 1995)

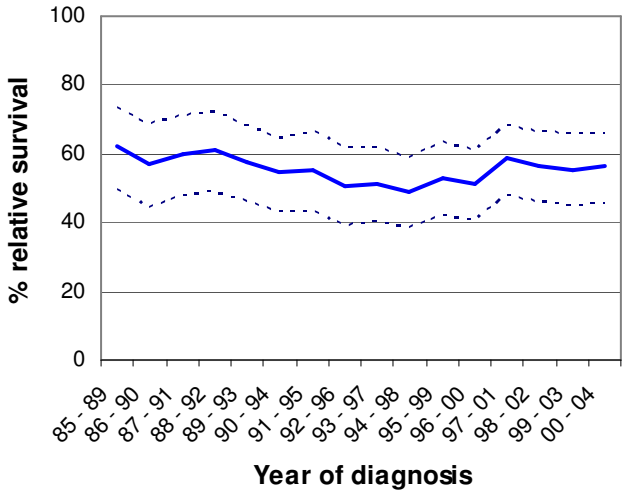
column and skull. 96% of chordomas diagnosed between 1985 and 2009 were within these regions. Chordomas arise in the notochord, so the tumours coded to the skull most probably actually relate to the base of skull. The small proportion of chordomas diagnosed in the bones of the upper limb most probably represents coding issues within the NCDR data.

Approximately 50% of chordomas diagnosed in patients aged 60-69 years (the largest cohort of chordoma patients) arise within the pelvic bones. Figure 4.4.1a also shows that chordomas diagnosed in patients under the age of 30 years appear to be confined to the bones of the skull and face, and to the vertebral column. However, this represents a very small proportion of patients with a chordoma; only 39 patients under the age of 30 years were diagnosed with a chordoma between 1985 and 2009.

4.4.3 Chordoma survival

Five-year relative survival rates for chordomas remained static at approximately 60% between 1985 and 2004, and may have even deteriorated prior to 2000. Survival rates could not be broken down further by anatomical sub-site due to the exceptionally small incidence rates involved

Figure 4.4.3: 5-year chordoma relative survival rates (England: 1985 – 2004)



Five-year chordoma relative survival rates are slightly lower than the 68% reported by McMaster et al. Interestingly, chordoma relative survival rates follow a similar pattern to those of Ewing's sarcoma, in that they appear to deteriorate in the 1990's and improve from 2000 onwards.

4.5 Ewing's Family of Tumours

The Ewing's family of tumours consists of M9260 (Ewing's sarcoma) and M9364 (peripheral neuroectodermal tumour). However, Ewing's sarcoma is by far the most common histological diagnosis, accounting for 97% of this classification of tumours. For the purposes of the following analyses, the variants are aggregated to produce incidence and survival rates.

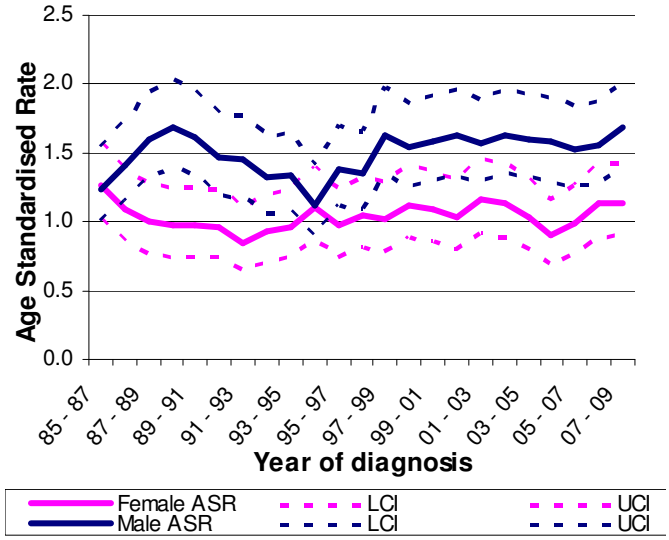
4.5.1 Incidence of Ewing's sarcoma

Out of the 9,574 primary bone tumours diagnosed between 1985 and 2009, 1,458 (15%) were either a Ewing's sarcoma or a primitive neuroectodermal tumour. Contrastingly, the WHO classification of bone tumours estimates that Ewing's sarcoma account for around 8% of primary bone malignancies. There were 62 new diagnoses of Ewing's sarcoma of the bone in 2009.

Figure 4.5.1a shows the three-year rolling incidence rates of patients diagnosed with a Ewing's sarcoma between 1985 and 2009. Rates are significantly higher in males (1.5 per million) than

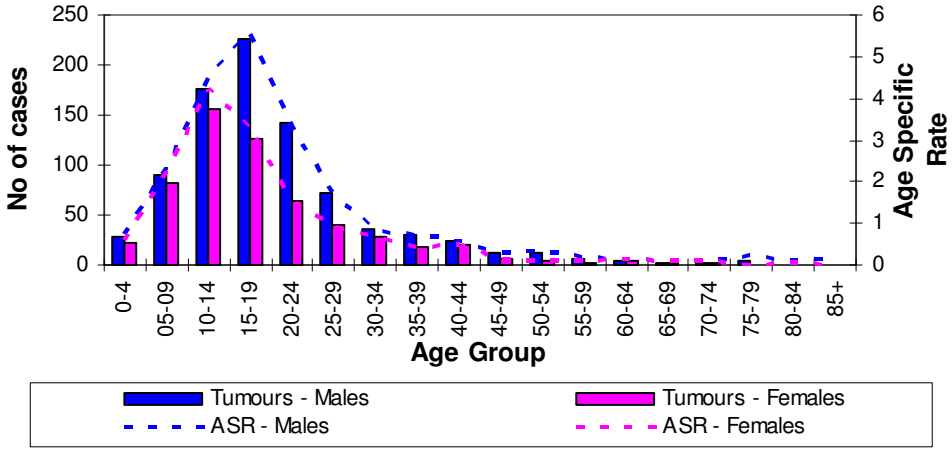
females (1 per million), although there appears to be a period in the mid 1990's where the rates were almost identical.

Figure 4.5.1a: 3-year rolling age standardised Ewing's sarcoma incidence rates by sex (England: 1985 – 2009)



Approximately 85% of Ewing's sarcomas are diagnosed in patients under the age of 30 years, and 35% of primary bone malignancies diagnosed in this age group are Ewing's sarcomas.

Figure 4.5.1a: Age standardised Ewing's sarcoma incidence rates by sex (England: 1985 – 2009)

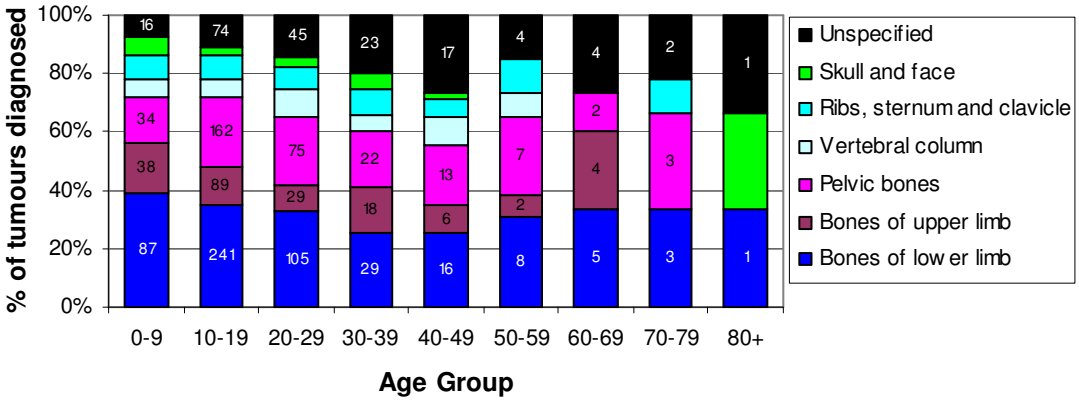


Ewing's sarcomas are exceptionally rare in the elderly. Only 117 Ewing's sarcomas were diagnosed in patients aged 40 years and over between 1985 and 2009. The highest age specific rate occurs in males aged between 15 and 19 years (5.7 per million), where the rate is over 1.7 times that of females in the same age group (3.2 per million).

4.5.2 Variation in Ewing's sarcoma incidence with anatomical site

Ewing's sarcoma can arise in any bone within the body. However, the most common sites for diagnosis are within the bones of the extremities (47%) and the pelvic bones (22%). Unlike osteosarcoma, the site of diagnosis does not generally appear to be dependent on age (Figure 4.5.2), but pelvic tumours become far more common in patients over the age of 10 years. The proportion of tumours with unspecified site of diagnosis is high across all age groups.

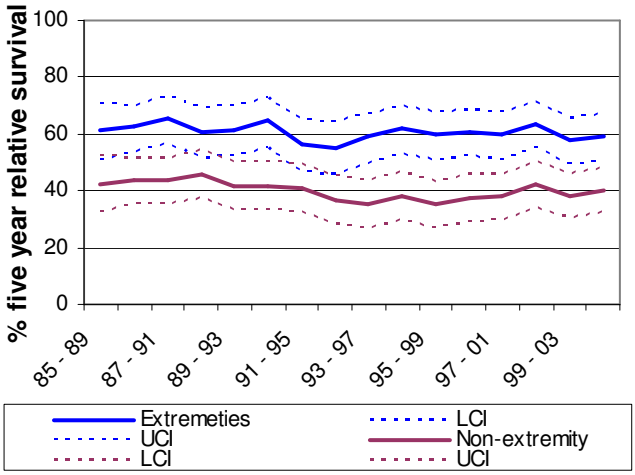
Figure 4.5.2: Proportion of Ewing's sarcomas diagnosed by age and anatomical site (England: 1985 – 2009)



4.5.3 Ewing's sarcoma survival

Ewing's sarcoma five-year relative survival rates remained static between 1985 and 2009 at around 50%, and may have actually deteriorated in the mid 1990's.

Figure 4.5.3: 5-year Ewing's sarcoma relative survival by anatomical sub-site (England: 1985 – 2004)



As with osteosarcomas and chondrosarcomas, 5-year Ewing's sarcoma relative survival rates are significantly higher in patients with extremity tumours than non-extremity tumours.