



Public Health
England

National Cancer Intelligence Network

Head and Neck Sarcoma of the Bone and Soft Tissue – Incidence, Survival and Surgical Treatment

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The intelligence networks

Public Health England operates a number of intelligence networks, which work with partners to develop world-class population health intelligence to help improve local, national and international public health systems.

National Cancer Intelligence Network

The National Cancer Intelligence Network (NCIN) is a UK-wide initiative, working to drive improvements in cancer awareness, prevention, diagnosis and clinical outcomes by improving and using the information collected about cancer patients for analysis, publication and research.

National Cardiovascular Intelligence Network

The National Cardiovascular Intelligence Network (NCVIN) analyses information and data and turns it into meaningful timely health intelligence for commissioners, policy makers, clinicians and health professionals to improve services and outcomes.

National Child and Maternal Health Intelligence Network

The National Child and Maternal Health Intelligence Networks (NCMHIN) provides information and intelligence to improve decision-making for high quality, cost effective services. Their work supports policy makers, commissioners, managers, regulators, and other health stakeholders working on children's, young people's and maternal health.

National Mental Health Intelligence Network

The National Mental Health Intelligence Network (NMHIN) is a single shared network in partnership with key stakeholder organisations. The Network seeks to put information and intelligence into the hands of decision makers to improve mental health and wellbeing.

National End of Life Care Intelligence Network

The National End of Life Care Intelligence Network (NEoLCIN) aims to improve the collection and analysis of information related to the quality, volume and costs of care provided by the NHS, social services and the third sector to adults approaching the end of life. This intelligence will help drive improvements in the quality and productivity of services.

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Executive summary

Bone and soft tissue sarcomas are a rare group of tumours which respectively account for 0.2% and 1% of all malignancies diagnosed.

With an incidence of approximately 38 tumours per annum, bone sarcomas of the skull and facial skeleton account for 9% of all bone sarcomas. Soft tissue sarcomas of the head and neck region are slightly more common with an annual incidence of 190 tumours and account for around 10% of all soft tissue sarcomas. Soft tissue sarcoma incidence rates are significantly higher in males than females, and the incidence of skin sarcomas is nearly three times as high in males as females.

Leiomyosarcoma (18%), angiosarcoma (12.5%) and rhabdomyosarcoma (13.6%) are the most common sarcoma morphological sub-types arising in the connective and soft tissue of the head and neck region. The latter two sub-types are highly aggressive tumours.

Five year relative survival rates for patients with a sarcoma of the head and neck region are around 73% for bone sarcoma and 64% for connective and soft tissue sarcoma.

Soft tissue sarcoma patients less than ten years of age are least likely to have any surgical intervention for their tumours, with a resection rate of around 35% for this age group. These patients tend to have a specific diagnosis of rhabdomyosarcoma and are treated with chemotherapy.

Following the publication of the Improving Outcomes Guidance for Patients with Head and Neck Cancer in 2004, referrals into large specialist centres which host head and neck cancer multi-disciplinary teams (MDTs) increased significantly between 2000-2004 and 2005-2010. However, 139 different hospital Trusts oversaw the surgical management of at least one head and neck sarcoma patient in the latter period, and 95 hospital Trusts had a surgical caseload of fewer than five patients over this period. Moreover, in the latter period, 8% of patients with a bone sarcoma were treated within a hospital Trust which did not host a head and neck cancer, sarcoma or brain and central nervous system MDT and was not a children's hospital.

Sarcomas of the head and neck region represent an important group of tumours as it is often difficult to obtain wide margins due to the anatomical location. This means the head and neck sarcomas are prone to recurrence and metastasis. Early diagnosis of these tumours is thus essential for improving outcomes.

Acknowledgements

The West Midlands Public Health England Knowledge and Intelligence Team would like to thank the South East Knowledge and Intelligence Team for the provision of the anatomical cancer site ICD-10 codes applied for the identification of head and neck cancer. We would also like to thank the National Cancer Intelligence Network Head and Neck Site Specific Clinical Reference Group, and in particular Mr Richard Wight and Mr Austen Smith for their helpful comments, and the London School of Hygiene and Tropical Medicine for providing the life tables used in this analysis.

Introduction

Sarcomas are a group of rare heterogeneous neoplasms which can arise in the bones or within the soft and connective tissue of the body. Bone sarcomas account for approximately 0.2% of malignant tumours diagnosed annually¹ whereas soft tissue sarcomas are slightly more common and represent approximately 1%². Over 80 different types of sarcoma were diagnosed in England between 1990 and 2000. The most common types of bone sarcoma are chondrosarcoma, osteosarcoma and Ewing's sarcoma. The most common types of soft tissue sarcoma are leiomyosarcoma, liposarcoma and sarcoma NOS².

It is estimated that 5% of bone sarcomas arise in the bones of the skull and face³, and that 10% of soft tissue sarcomas arise in the soft tissue of the head and neck region⁴. Due to the diverse morphological sub-types and their rarity, treatment of sarcomas arising in the head and neck region is challenging. The Improving Outcomes Guidance (IOG) on the Diagnosis and Treatment of Head and Neck Cancers was published in 2004⁵. This recommended that head and neck cancers should be treated by specialised surgical and oncological teams. The Improving Outcomes Guidance for Bone and Soft Tissue Sarcoma which was published in 2006 advised that any patient with a diagnosis of bone or soft tissue sarcoma should have their care overseen by a sarcoma multi-disciplinary team (MDT)⁶. The IOG also advised that, for patients with site specific sarcomas such as gynaecological or head and neck tumours, there should be an established relationship between the sarcoma and site specific MDTs.

Information on head and neck sarcoma incidence, outcomes and treatment is currently very limited and is generally based on small cohorts of patients recorded in hospital treatment databases. The aim of this report is to establish the incidence and survival rates for patients diagnosed with bone and soft tissue sarcomas arising in the head and neck region based on English cancer registration data and, more importantly, to establish where patients with these rare sarcomas were surgically treated in the period 1990-2010.

Methods

The Public Health England West Midlands Knowledge and Intelligence Team (WMKIT) is the National Cancer Intelligence Network (NCIN) national lead analytical team for bone and soft tissue sarcoma. The South East Knowledge and Intelligence Team is the national lead for head and neck cancer. The lead KITs analyse national data on the incidence, mortality, survival and treatment of their respective specific cancer site(s) in England. These analyses are usually conducted using the National Cancer Data Repository (NCDR), an evolving source of data collected by the eight regional cancer registration offices which covers all cancers

diagnosed in England. The current version of the NCDR includes all malignancies diagnosed in England between 1990 and 2010.

Soft tissue sarcomas are classified by both a 10th revision of the International Classification of Diseases (ICD-10) site code and an ICD-O3 morphology code. Within the ICD-10 coding system, the prefix 'C' locates the code within the 'neoplasm', or cancer, subgroup, and the following numbers 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-13' denotes a malignant neoplasm of the hypopharynx, and 'C-131' represents a malignant neoplasm of the aryepiglottic fold within the hypopharynx. Tumours arising within the bones and soft tissue of the head and neck region are identified using the ICD-10 codes in Appendix A and the ICD-O3 codes in Appendix B.

The Hospital Episode Statistics (HES) dataset to which the KITs have access contains all inpatient and day case patients who have at least one admission where any cancer (benign or malignant) is recorded. The HES dataset records information such as the hospital of treatment or care, the type of surgical treatment provided, and other diagnoses observed in the patient. Primary surgical treatment for a head and neck cancer was identified within the HES data as any curative surgical procedure (as defined by the UK and Ireland Association of Cancer Registries: UKIACR) to the head and neck undertaken within six months of initial diagnosis (Appendix C). Consultant specialist codes recorded in HES were used to establish the speciality of the corresponding consultant overseeing the patient's care during each hospital admission.

Age-standardised (ASR) and age specific (ASIR) incidence rates are expressed as numbers per million population throughout. 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 strsr programme which calculates relative survival estimates using the Ederer II method. Five-year relative survival was calculated using 5-year rolling averages. National life tables were obtained from the Cancer Research UK Cancer Survival Group at the London School of Hygiene and Tropical Medicine.

Tables containing the data used to construct the figures in this reports are provided in Appendix D.

Incidence

Between 1990 and 2010, 4,796 patients were diagnosed with a sarcoma arising in the head and neck region, of which 793 arose in the bones of the skull and face, and 4,003 within the connective and soft tissue. Connective tissue sarcomas were the most common type of head and neck sarcoma in males and females (45.1% and 36.0% respectively) (Figure 1). Bone sarcomas constituted 20.9 % of all head and neck sarcomas in females and 14.3% in males. Skin sarcomas formed a higher proportion of all head and neck sarcomas in males (19.4% and 13.4% respectively).

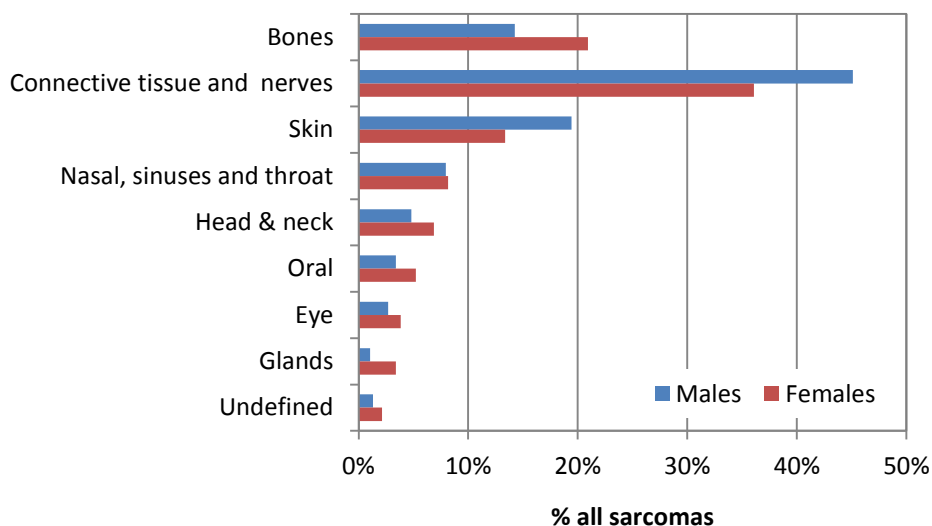


Figure 1: Head and neck sarcomas in males and females (England: 1990-2010)

The overall number of sarcomas diagnosed in males exceeded those diagnosed in females by a ratio of 1.9 to 1. The highest male to female incidence ratios were seen for skin tumours (2.78 to 1.00) and connective tissue and nerves (2.40 to 1.00) (Figure 2). The number of tumours diagnosed in glands was higher in females although this is based on 89 sarcomas diagnosed between 1990 and 2010.

Male to female ratios for bone sarcoma increased gradually from 1.15 in patients aged 0-9 years to 1.95 in patients aged 60-69 years, and then fell below 1.00 in those aged 70 years and above (Figure 3). Male to female ratios for soft tissue sarcomas increased rapidly from 1.61 at the age of 50-59 years to 3.17 at the age of 70-79 years.

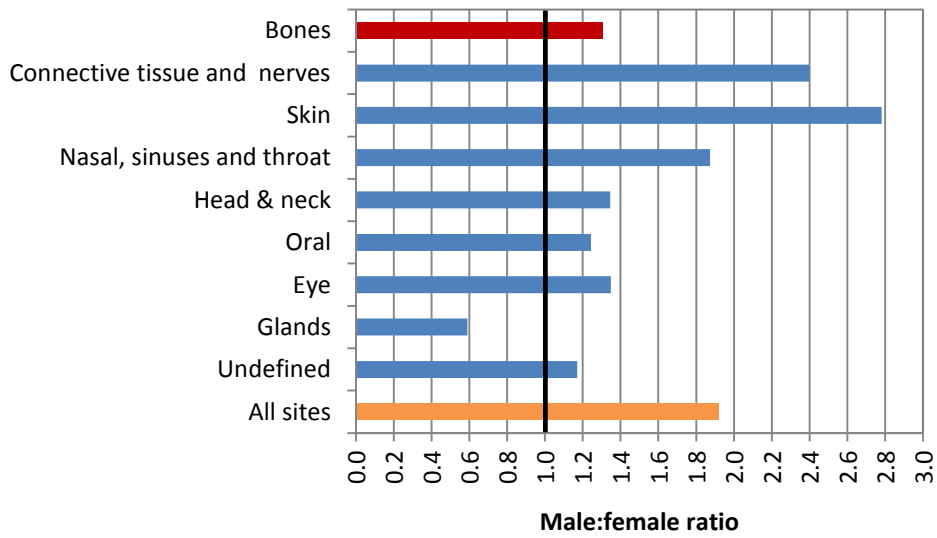


Figure 2: Head and neck sarcomas male to female ratios (England: 1990-2010)

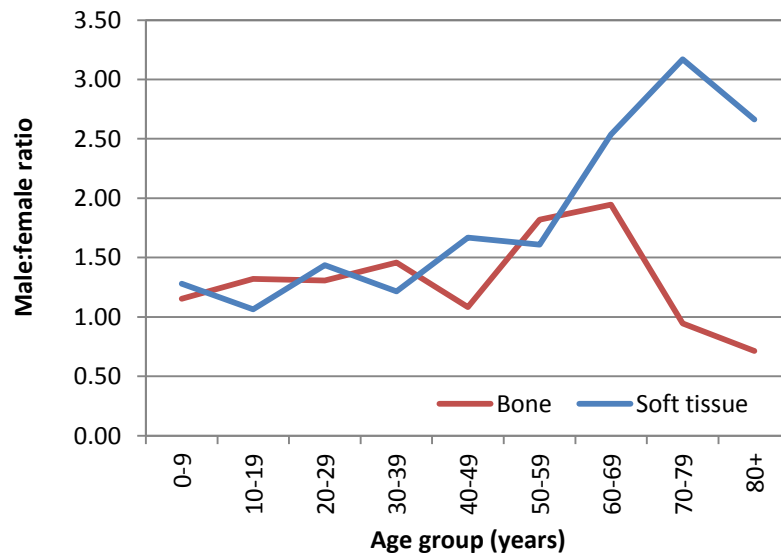


Figure 3: Head and neck bone and soft tissue sarcoma male to female ratios (England: 1990-2010)

Bone Sarcomas of the Skull and Facial Skeleton

Between 1990 and 2010 there were 793 new diagnoses of sarcomas arising in the bones of the skull and facial skeleton. On average, 38 (range 19 to 59) bone sarcomas of the skull and facial skeleton were diagnosed annually, accounting for 10% of all bone sarcomas. There were no significant differences in the male (0.8 per million) and female (0.6 per million) age-standardised incidence rates over the period 1990-2010 (Figure 4a). However, while female age-standardised incidence rates varied little between 1990 and 2010, male age-standardised rates in 2005-2010 were 1.5 times as high as those in 1993-2004. This difference is statistically significant.

Age-specific incidence rates for bone sarcomas of the skull and facial skeleton did not follow the bimodal incidence pattern which is normally associated with bone sarcomas, but increased gradually with age, from 0.3 per million in the youngest age group (0-4 years) to 1.9 per million in the 80-84 year age group (Figure 4b). Between the ages of 50 and 74 years, age-specific incidence rates in males exceeded those in females by a ratio of 1.8:1 and this difference is statistically significant.

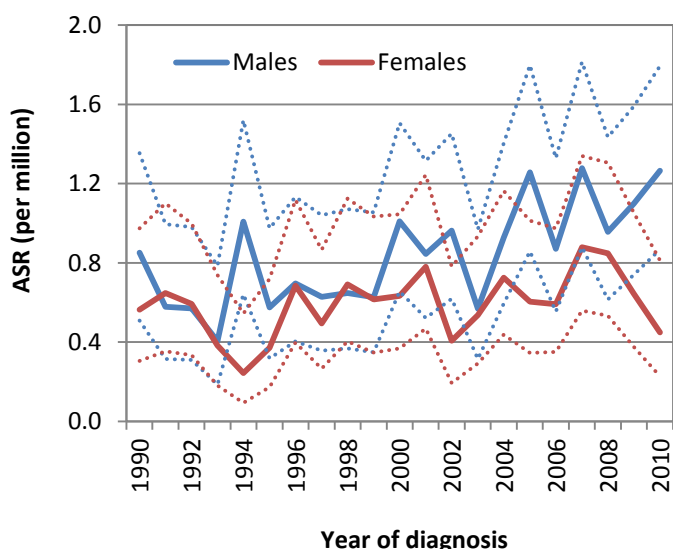


Figure 4a: Skull and facial bone sarcoma age-standardised incidence rates (England: 1990-2010)

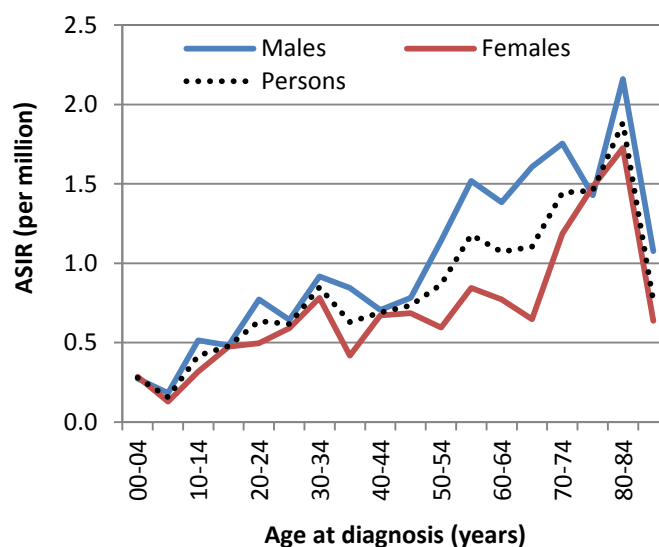


Figure 4b: Skull and facial bone sarcoma age-specific incidence rates (England: 1990-2010)

The most common histological diagnoses in the bones of the skull and facial skeleton were chondrosarcoma (232, 29%), osteosarcoma (190, 24%) and chordoma (129, 16%). Less common variants of bone sarcoma such as ameloblastoma and odontogenic tumours accounted for 10% and 5% of bone sarcomas respectively. Chondrosarcomas accounted for a higher proportion of all bone sarcomas in females (37% compared with 23% in males).

Sarcomas of the Connective and Soft Tissue

Incidence rates

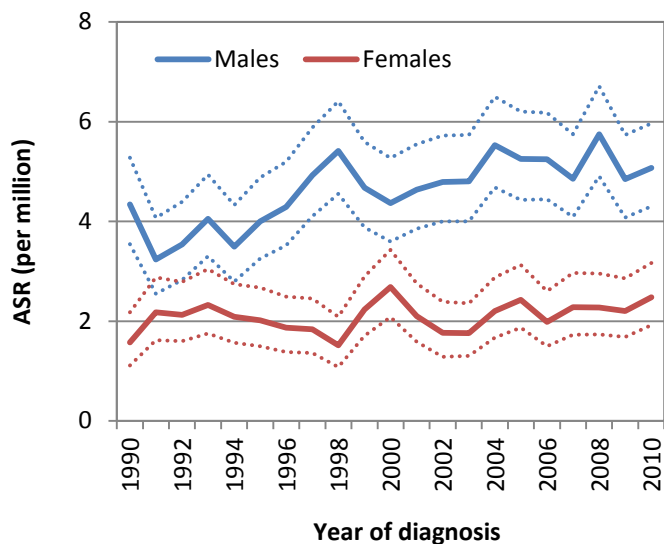


Figure 5a: Head and neck soft tissue sarcoma age-standardised incidence rates (England: 1990-2010)

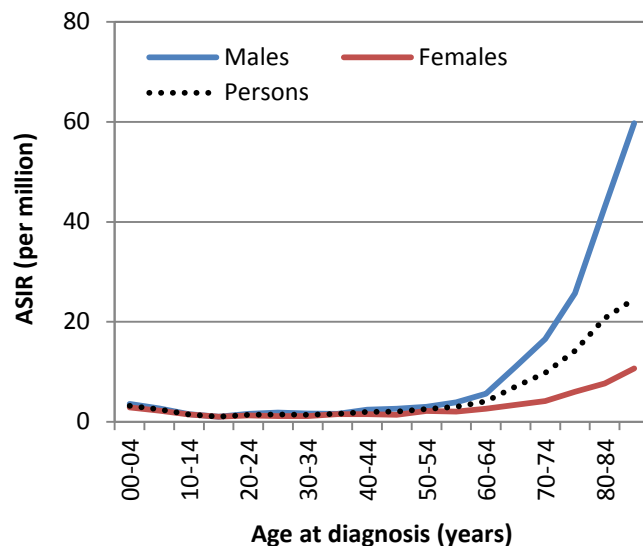


Figure 5b: Head and neck soft tissue sarcoma age-specific incidence rates (England: 1990-2010)

Anatomical sites

During the twenty-one year period studied, there were 4,003 new diagnoses of soft tissue sarcoma arising in the head and neck region. On average there were around 190 head and neck soft tissue sarcomas diagnosed annually, accounting for 9% of all soft tissue sarcomas. The age-standardised incidence rates for all persons over the period 1990-2010, oscillated around 3.5 per million population. Incidence rates in males (4 - 5 per million) were higher than those in females (2 per million) (Figure 5a). This difference is statistically significant from 1995 onwards. Significantly higher incidence rates in male head and neck patients have been found in other reviews, but there are no clear explanations for this finding⁴. Male age-standardised incidence rates increased significantly between 1993-1998 and 2005-2010 from 4.4 per million to 5.2 per million, while female incidence rates remained relatively constant. Age-specific incidence rates increased rapidly with age from the age of 65 years onwards and were significantly higher in males than in females. In males aged 85 years and over, the incidence rate (60 per million) exceeded that in females (10.7 per million) by a ratio of 5:1 (Figure 5b).

Table 1: Soft Tissue sarcomas of the head and neck region: anatomical sites of diagnosis (England: 1990-2010)

ICD-10 code	Anatomical site	No. cases	%
C47, C49	Connective tissue, nerves	2015	50.3%
C44	Skin	832	20.8%
C30-C33	Nasal, sinuses, throat	385	9.6%
C07-C14	Head and neck	265	6.6%
C00-C06	Oral	193	4.8%
C69	Eye	148	3.7%
C73, C74	Glands	89	2.2%
C76	Undefined	76	1.9%
	All sites	4,003	100.0%

Table 1 shows the number of head and neck soft tissue sarcomas diagnosed at each anatomical site and the ICD-10 codes used to identify these sites. Head and neck sarcomas occurred most commonly in connective and soft tissue (51%) and skin (21%). Increases in the numbers of sarcomas diagnosed at these two sites accounted for most of the increase in male soft tissue sarcoma age-standardised incidence rates seen between 1993-98 and 2005-10 (Figure 6).

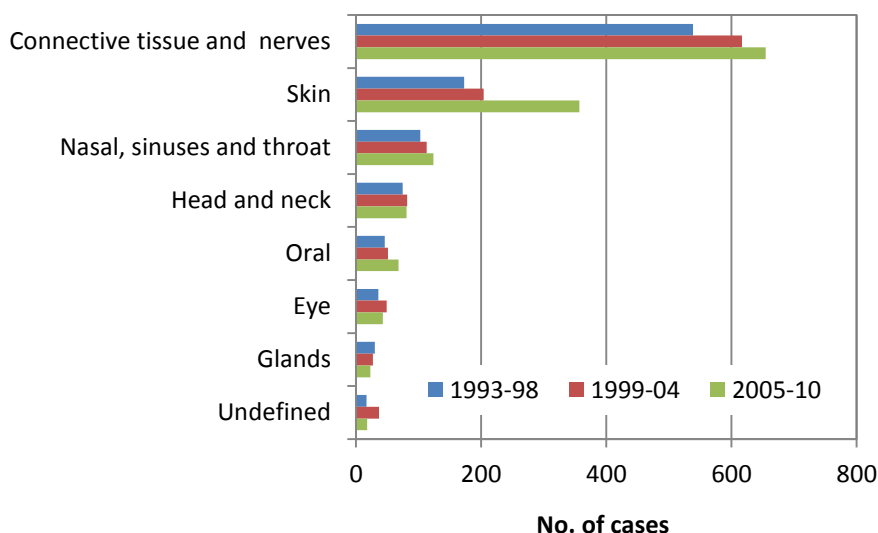


Figure 6: Soft Tissue sarcomas of the head and neck region: numbers of cases diagnosed in each anatomical site in 1993-98, 1999-04 and 2005-10

Morphological sub-types

Table 2 shows the number of head and neck soft tissue sarcomas in each morphological sub-type in each anatomical site. The most common head and neck soft tissue sarcoma morphological sub-types were leiomyosarcoma (724, 18%), sarcoma NOS (663, 17%) and rhabdomyosarcoma (545, 14%). This figure for rhabdomyosarcoma is well within the range normally found, but the proportion of leiomyosarcomas is much higher than generally cited (5%)⁷.

Table 2: Soft Tissue sarcomas of the head and neck region: anatomical sites of diagnosis and morphological sub-types (England: 1990-2010)

Histology	Connective tissue, nerves	Skin	Nasal, sinuses and throat	Head and neck	Oral	Eye	Glands	Undefined	Total
Leiomyosarcoma	432	158	54	25	31	4	15	5	724
Angiosarcoma	319	128	17	11	10	3	7	7	502
Rhabdomyosarcoma	164	9	100	98	55	106	2	11	545
Chondrosarcoma	15	0	67	0	0	1	0	3	86
Dermatofibrosarcoma	1	273	0	0	0	0	0	2	276
Fibrosarcoma	38	12	9	3	3	3	5	1	74
Liposarcoma	160	8	4	19	20	2	2	4	219
MFH	231	122	11	11	7	2	2	1	387
MPNST	171	4	3	3	2	1	0	2	186
Sarcoma, NOS	338	108	76	42	47	12	25	15	663
Other	146	10	44	53	18	14	31	25	341
Chordoma	2	0	18	14	0	5	17	2	58
Ewing's sarcoma	20	2	4	3	3	2	6	7	47
Myxofibrosarcoma	32	5	6	3	1	1	1	1	50
Osteosarcoma	0	0	2	0	0	2	0	1	5
Other	44	3	13	28	13	4	6	11	122
Synovial	48	0	1	5	1	0	1	3	59
Total	2015	832	385	265	193	148	89	76	4003

MFH= Malignant Fibrous Histiocytoma

MPNST= Malignant peripheral nerve sheath tumour

Figure 7 shows the main anatomical sites associated with each morphological sub-type. Dermatofibrosarcomas occurred predominantly in skin (98.9%) and chondrosarcomas in the nasal cavities, sinuses and throat (77.9%). Skin was also a relatively common site for malignant fibrous histiocytoma (MFH) (31.5%), angiosarcoma (25.5%) and leiomyosarcoma (21.8%).

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

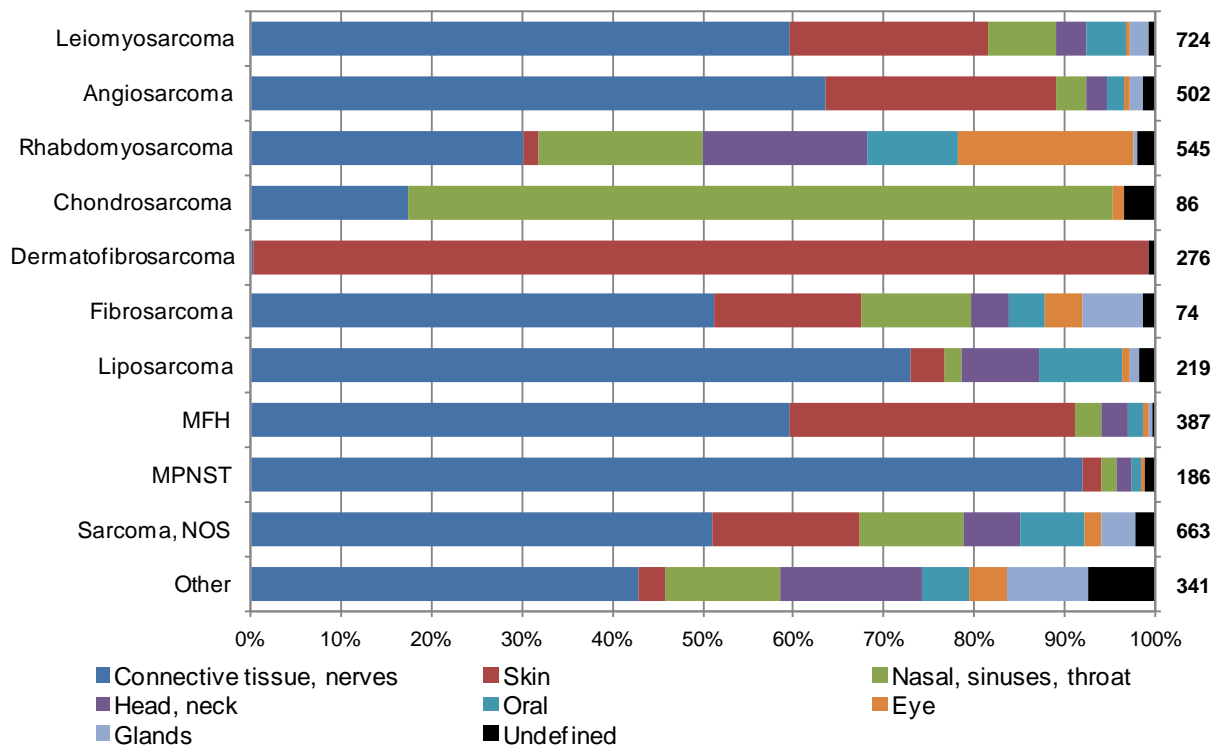


Figure 7: Soft Tissue sarcomas of the head and neck region: anatomical sites of diagnosis and morphological sub-types (England: 1990-2010)

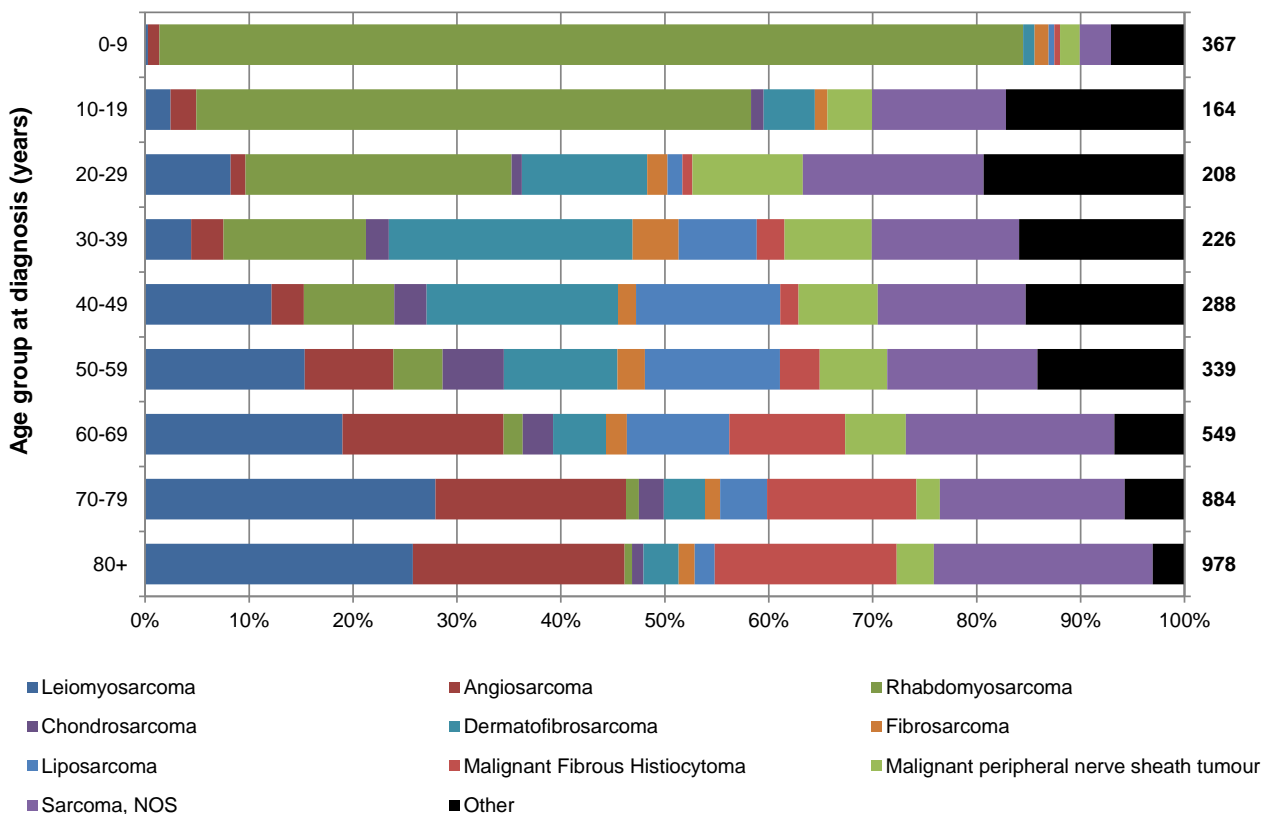


Figure 8: Soft tissue sarcomas of the head and neck region – most common morphological sub-types in each age group (England: 1990-2010)

Head and neck soft tissue sarcoma morphological subtypes varied with age at diagnosis (Figure 8). Leiomyosarcomas were rarely found in patients aged less than 40 years. Rhabdomyosarcomas (RMS) were the most predominant diagnosis in patients aged less than 10 years, accounting for 83% of diagnoses in this age group (56% of all head and neck rhabdomyosarcomas were diagnosed in the youngest patients). The majority of head and neck angiosarcomas (89%) and malignant fibrous histiocytoma (MFH) (93%) were diagnosed in patients aged 60 years and over. Dermatofibrosarcomas were most likely to be diagnosed in patients between the ages of 30 and 50 years.

Survival

Relative survival rates for patients with bone or soft tissue (STS) head and neck sarcomas diagnosed in 2001-2005 are shown in Figure 9a. Patients with a soft tissue head and neck sarcoma had a 1-year relative survival rate of 84% and a 5-year relative survival rate of 64%. Bone head and neck sarcoma patients diagnosed in the same period had a 1-year relative survival rate of 90% and a 5-year relative survival rate of 73%. Although patients with a bone head and neck sarcoma had higher survival rates, these differences are not statistically significant.

There were no significant effects of sex or age at diagnosis on the survival of head and neck bone or soft tissue sarcoma patients, although males and younger people did have slightly higher survival. There was, however, a significant difference in relative survival between soft tissue sarcoma patients diagnosed with a sarcoma of the skin of the head and neck compared to the other soft tissues. Patients who were diagnosed with a skin sarcoma had an 85% 5-year relative survival rate, compared to the 60% 5-year relative survival rate in patients diagnosed with a soft tissue sarcoma elsewhere in the head and neck region (Figure 9a).

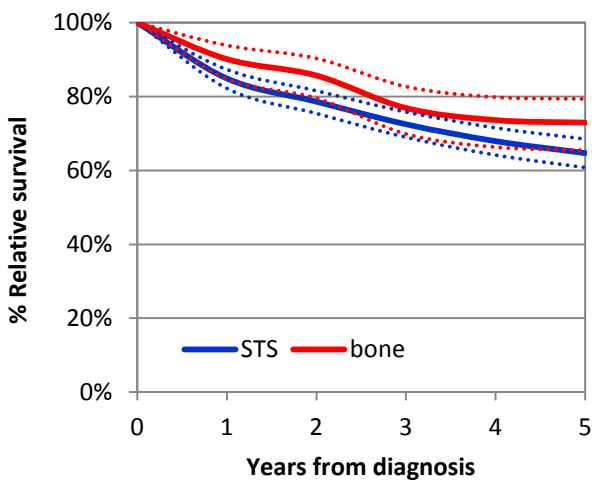


Figure 9a: Head and neck bone and soft tissue sarcoma 1-5 year relative survival (England, 2001-2005)

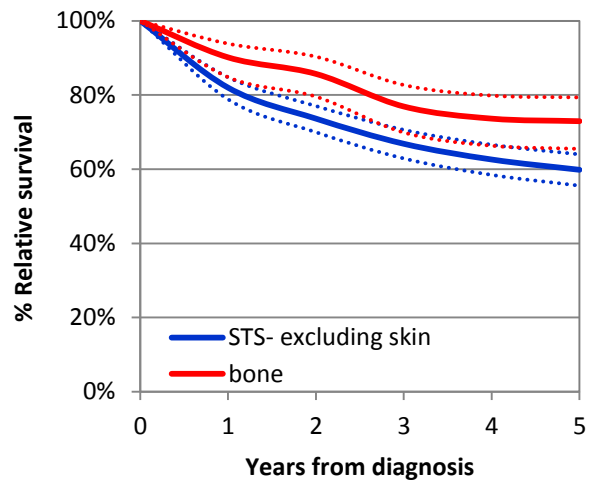


Figure 9b: Head and neck bone and soft tissue sarcoma 1-5 year relative survival (England, 2001-2005)

If soft tissue sarcomas of the skin are excluded, 5-year relative survival rates for patients with soft tissue sarcomas of the head and neck region (60%) are significantly worse than for those with sarcomas arising in the bones of the skull and facial skeleton (73%) (Figure 9b). Although exceptionally rare, 5-year relative survival rates were lowest for patients with angiosarcomas arising in the soft tissue of the head and neck (31%) and for alveolar rhabdomyosarcomas (42%).

Surgical treatment

The inpatient/day case HES data were analysed to identify surgery related admissions for patients diagnosed with a sarcoma arising within the bone or soft tissue of the head and neck region. Although inpatient/day case HES started between 1997 and 1998, to ensure that the most complete data were available, the diagnosis years were limited to the period 2000 to 2010, during which time 2,842 tumours were diagnosed: 2,351 soft tissue sarcomas of the head and neck, and 491 bone sarcomas of the skull and facial skeleton.

Bone Sarcomas of the Skull and Facial Skeleton

Of the 491 bone, skull and facial sarcomas diagnosed between 2000 and 2010, a HES record relating to curative surgical treatment was identified for 329 (67%), and 469 curative surgical operations were performed during 386 hospital admissions. A small number of patients (13, 4%) were treated in more than one hospital Trust during the six month period following their diagnosis. The most commonly performed curative surgical procedures were extirpation of lesion of cranium, and full and partial excision of the mandible. Because of the small number of head and neck bone sarcomas diagnosed, and the lack of detailed staging information, it is not possible to examine long term outcomes based on the hospital Trust of surgical treatment.

Patients 80 years and over were less likely to be treated surgically, with only 15 out of 36 patients (42%) having surgical treatment (Figure 10). Of the 21 patients not treated surgically, the histological diagnoses were predominantly chordoma and variants of osteosarcoma. Factors such as co-morbidity and stage at diagnosis require investigation to establish why these older patients were not treated surgically and whether they received radiotherapy and/or chemotherapy.

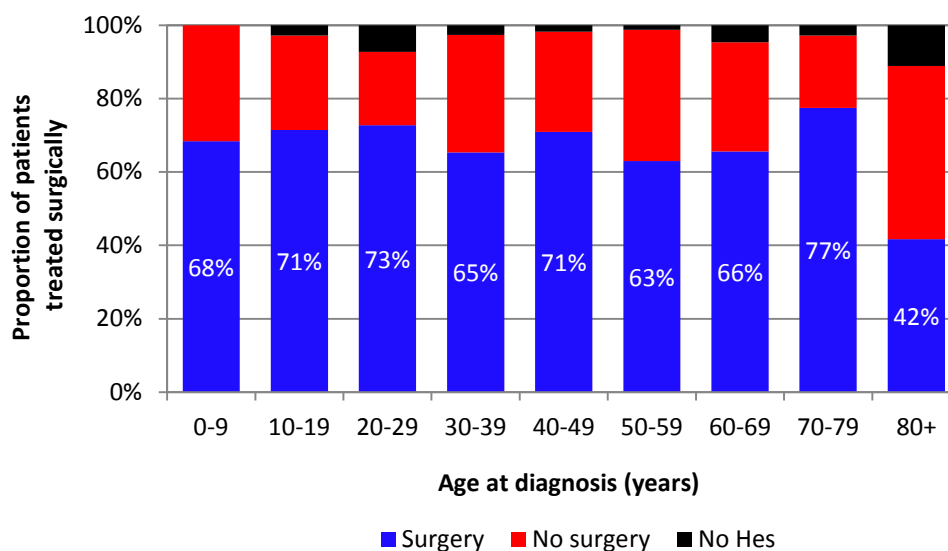


Figure 10: Variation with age in the proportion of skull and facial skeletal bone sarcoma patients treated with a curative surgical procedure (England: 2000-2010)

The numbers and proportion of patients receiving curative surgery for bone sarcomas of the head and neck region increased from 64.8% (127/196 patients) in 2000-2004 to 68.5% (202/295 patients) in 2005-2010. This difference is not statistically different ($p=0.4$).

With the information currently available, it is not possible to identify whether patients with bone sarcomas of the head and neck are discussed within a hospital hosting a sarcoma MDT. However, it is possible to identify the nature of the MDTs hosted by the hospital Trusts where patients received their surgical treatment.

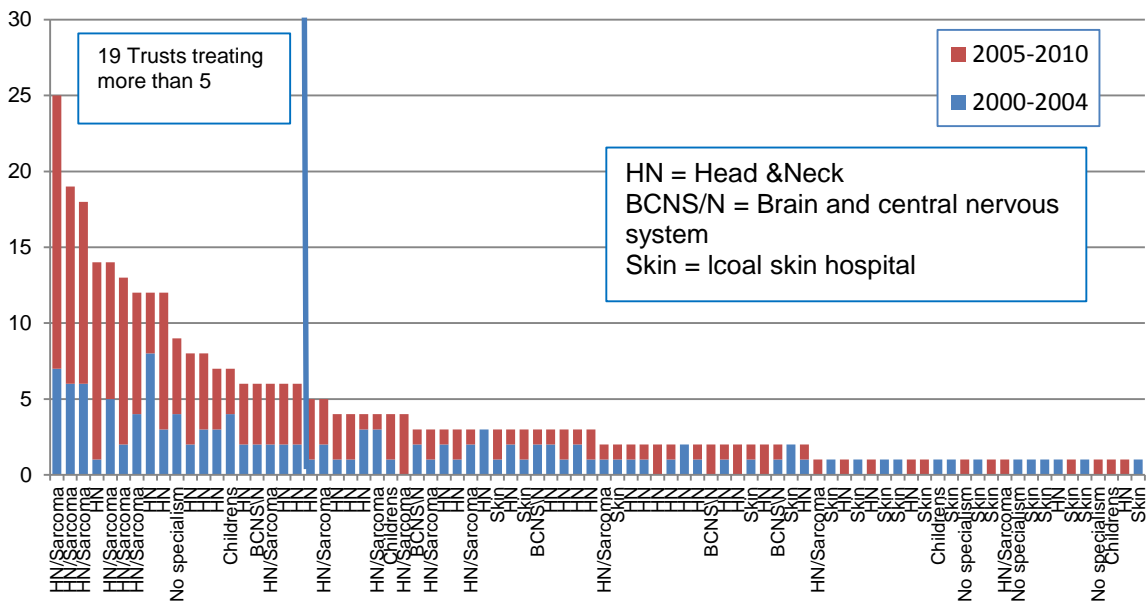


Figure 11: Number of bone sarcomas of the skull and facial skeleton treated within each NHS hospital Trust (England 2000-2010)

Figure 11 shows how the number of bone sarcomas of the skull and facial skeleton treated within each NHS hospital Trust in England in 2000-2004 and 2005-2010 varied with the nature of the MDT hosted by that Trust. Figure 12 summarises these data for the main types of MDT.

During the 11-year period studied, although 82 different hospital Trusts were identified to have treated at least one bone sarcoma of the skull and facial skeleton, 208 (61%) of the curative surgical operations were performed within the 19 Trusts which treated more than 5 bone sarcomas in total. In 2000-2004 65 different Trusts treated sarcomas of the head and neck region, and 68 (52%) of curative surgical operations were undertaken within the 19 Trusts which treated more than 5 bone sarcomas in the 11-year period studied. In 2005-2010, 66 different Trusts treated bone sarcomas of the head and neck region, and 140 (66%) curative surgical operations were undertaken with the 19 Trusts with the highest overall caseload. These differences are significantly different ($p=0.0098$). In the most recent data based on patients diagnosed with a bone sarcoma in 2010, 23 different hospital Trusts provided surgical treatment for at least one patient with a bone sarcoma arising in the skull or facial skeleton.

In 2000-2010, of the 386 hospital admissions relating to surgical treatment for a bone sarcoma arising in the skull or facial skeleton, 308 (80%) were within a hospital Trust hosting a head and neck cancer MDT; 164 (42%) of surgical procedures were undertaken within hospital Trusts hosting a head and neck cancer MDT only, with a further 144 (37%) in a hospital Trust hosting both head and neck cancer and sarcoma MDTs (Figure 12). The proportion of bone sarcomas treated in hospital Trusts with a head and neck, or sarcoma and head and neck MDT, increased from 73% in 2000-2004 to 84% in 2005-2010 (p=0.006), presumably as a result of the reorganisation of services which followed the publication of the corresponding head and neck cancer⁵ and sarcoma IOGs⁶.

The Trust with the largest caseload (which hosts a head and neck and sarcoma MDT) oversaw the curative surgical treatment of 25 tumours over the 11-year period (around two tumours annually). 8.3% of curative surgical operations (32 in total) were performed in a hospital Trust with a local skin cancer MDT, and a further 4% (15 in total) were provided in a children’s hospital, for which the average age at diagnosis was 5 years (range 0 to 12 years). The curative surgical procedures undertaken within hospital Trusts hosting brain and central nervous system/neuroscience” MDTs were related to the treatment of the cranium or excision of lesion of brain (8/21), with the remaining surgery codes relating to the treatment for tumours of the facial skeletal bones.

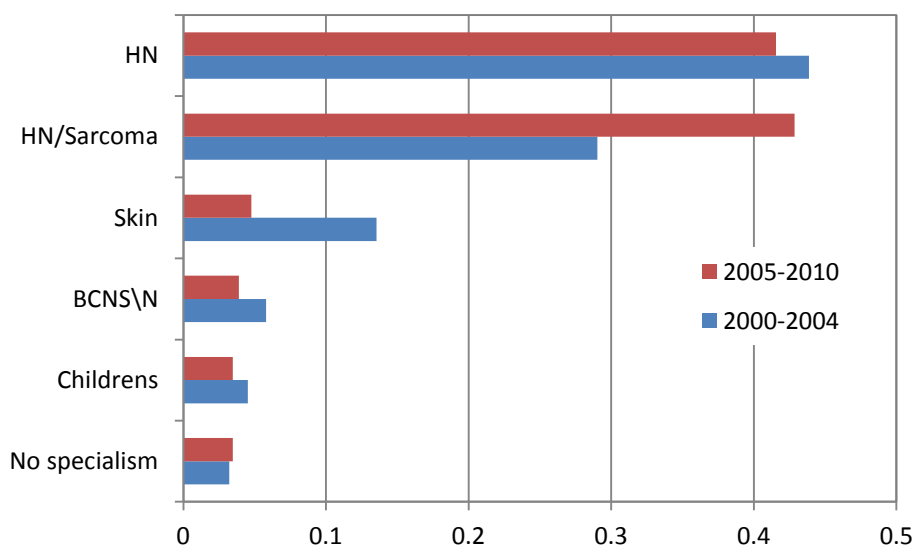


Figure 12: Number of skull and facial bone sarcomas treated within hospital Trusts with MDT services available (England 2000-2010)

HES data record the specialty of the consultant overseeing the care of patients during any particular episode. The surgical management of patients with bone sarcomas of the skull and facial skeleton was predominantly overseen by consultants specialising in oral and maxillofacial

surgery (46%), neurosurgery (31%) or by ear, throat and nose (ENT) specialists (15%) (Figure 13).

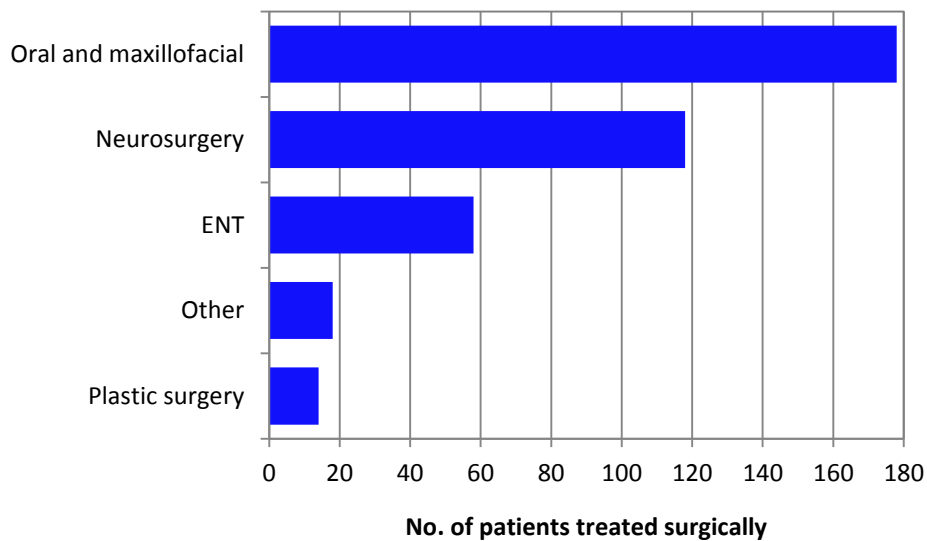


Figure 13: Number of skull and facial bone sarcomas treated by each consultant specialty (England: 2000-2010)

Sarcomas of the Connective and Soft Tissue

The Improving Outcomes Guidance for People with Sarcoma stipulates that more than one MDT may be required to discuss patients with head and neck sarcoma. The guidance also highlights the issue of the large number of benign tumours which cannot be distinguished from malignant tumours on the basis of clinical investigations alone.

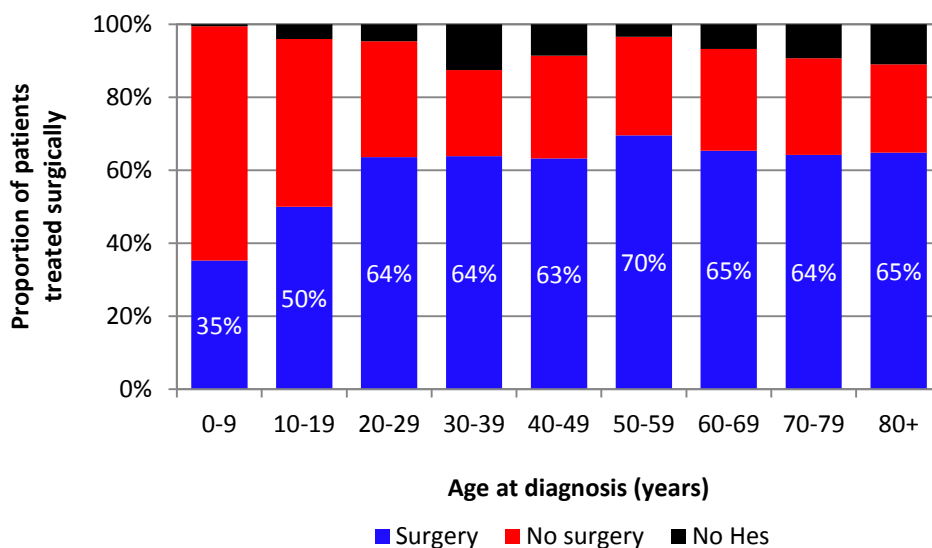


Figure 14: Proportion of patients with head and neck soft tissue sarcomas treated with a curative surgical procedure (England 2000-2010)

The overall proportion of patients receiving curative surgical treatment increased significantly between 2000-2004 and 2005-2010, from 59% to 65% [p=0.004]. Specifically, this is true of patients with a diagnosis of rhabdomyosarcoma (increased from 26% to 39%, p=0.021), sarcoma NOS (56% to 68%, p=0.017), and patients aged 0-9 years (26% to 42%, p= 0.023), 20-29 (54% to 69%, not significant) and 70-79 years (60% to 67%, not significant). Moreover, the proportion of patients aged 80 years and over who were not admitted to hospital decreased significantly from 14% to 9%.

Of the 2,351 patients diagnosed with a head and neck sarcoma between 2000 and 2010, a HES admission relating to curative surgical treatment was identified for 1,447 (62%). 107 patients received a curative surgical procedure in more than one hospital Trust. The majority of surgical treatment relating to the soft tissue of the head and neck region was recorded as “excision of lesion of skin of head and neck” (52%), with a further 9% relating to the “excision of lesion of soft tissue” or muscle. The remaining surgical treatments relate to specific areas of the head and neck region such as excision of lesion of internal nose, excision of lesion of tongue, excision of parotid gland etc.

Patients less than 10 years of age were least likely to be treated surgically, with only 35% of patients in this age group receiving surgery (Figure 14). The Intergroup Rhabdomyosarcoma Study Group recommended that children and adolescents should only be treated surgically provided that the surgery was not mutilating or cosmetically damaging⁸. In a study of 69 children with rhabdomyosarcomas arising in the soft tissue of the head and neck, only 12 (17%) were treated through surgical resection⁹. The preferred method for treating these tumours in younger patients is with radiotherapy and adjuvant chemotherapy. From the age of 20 years onwards, the proportion of patients receiving curative surgical procedures was uniform, with a resection rate of around 65%. Further investigations are required to understand the treatment pathways for patients who are not treated surgically, and to investigate the role of radiotherapy and chemotherapy for patients with these tumours.

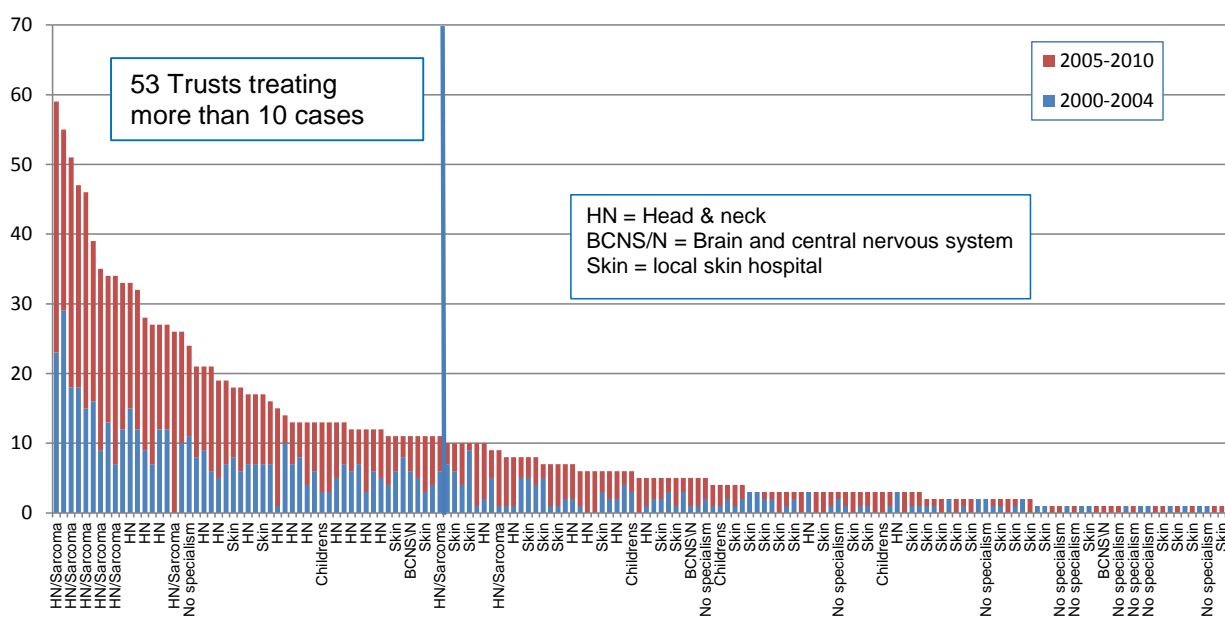


Figure 15: Number of head and neck soft tissue sarcomas treated in each hospital Trust (England: 2000-2010)

The 1,447 patients treated surgically had a total of 1,902 HES admissions relating to surgery. Figure 15 shows how the number of soft tissue sarcomas of the head and neck treated within each NHS hospital Trust in England from 2000-2010 varied with the nature of the MDT hosted by that Trust. Figure 16 summarises these data for the main types of MDT.

During the 11-year period 2000-2010, 159 different hospital Trusts were identified as providing surgical treatment for a head and neck soft tissue sarcoma. In 2000-2004 129 different Trusts treated soft tissue sarcomas of the head and neck region, and 447 (74%) of curative surgical operations were undertaken within the 53 Trusts which treated more than 10 soft tissue sarcomas in the 11-year period studied. In 2005-2010, 139 different Trusts treated soft tissue sarcomas of the head and neck region, and 703 (75%) (703) of curative surgical operations were undertaken within the 53 Trusts with the highest overall caseload. The three hospital Trusts with the highest surgical caseload all hosted a head and neck and sarcoma MDT.

Figure 16 shows that 1,290 (67%) of curative surgical procedures were undertaken in hospital Trusts with a head and neck cancer MDT and 649 (34%) in Trusts hosting both a head and neck and a soft tissue sarcoma MDT. A further 413 (22%) curative surgical procedures were undertaken in a Trust hosting a skin cancer MDT only, and 31 surgical operations on head and neck soft tissue sarcomas (<1%) were undertaken within a children’s hospital. The average age at diagnosis of these patients was 6 years (range 0 to 17 years). The curative surgical treatment undertaken for skin cancer in Trusts which collectively undertook 441 different curative surgical procedures, was predominantly skin related surgery (i.e. excision of lesion of head or neck, [257, 71%]) or related to excision of lesions of the ear, nose or mouth (46, 10%), although a further 32 (7%) procedures related to the excision of bone or soft tissue of the head and neck region.

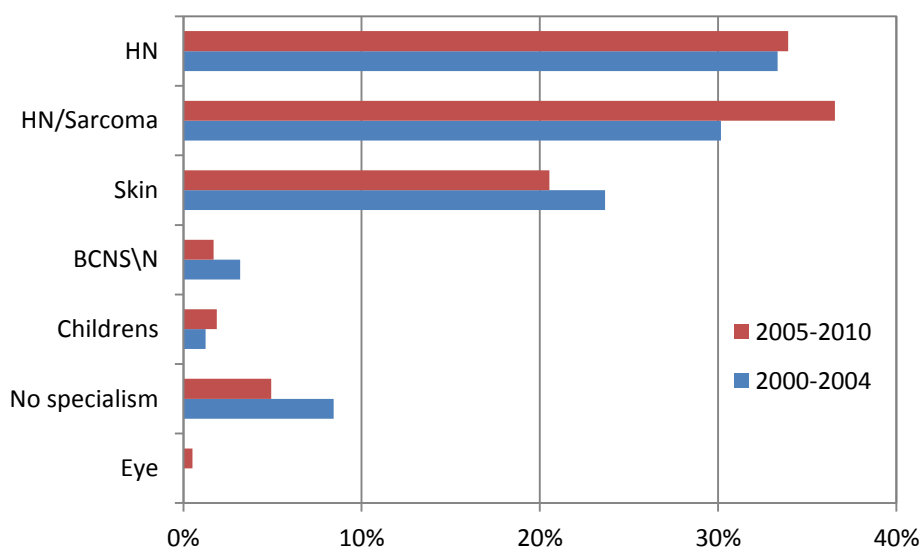


Figure 16: Number of head and neck soft tissue sarcomas treated within hospital Trusts with MDT services available (England 2000-2010)

The proportion of curative surgical procedures undertaken within hospital Trusts hosting a skin cancer MDT decreased significantly between 2000-2004 and 2005-2010 from 171 (24%) to 21%. The proportion of curative surgical procedures undertaken within a hospital hosting a head and neck cancer MDT increased significantly from 63% to 70% [p=0.015]. The HES data require further investigation to identify surgical treatment more than six-months after diagnosis to establish if any of these patients were later admitted into either a head and neck or sarcoma Trust.

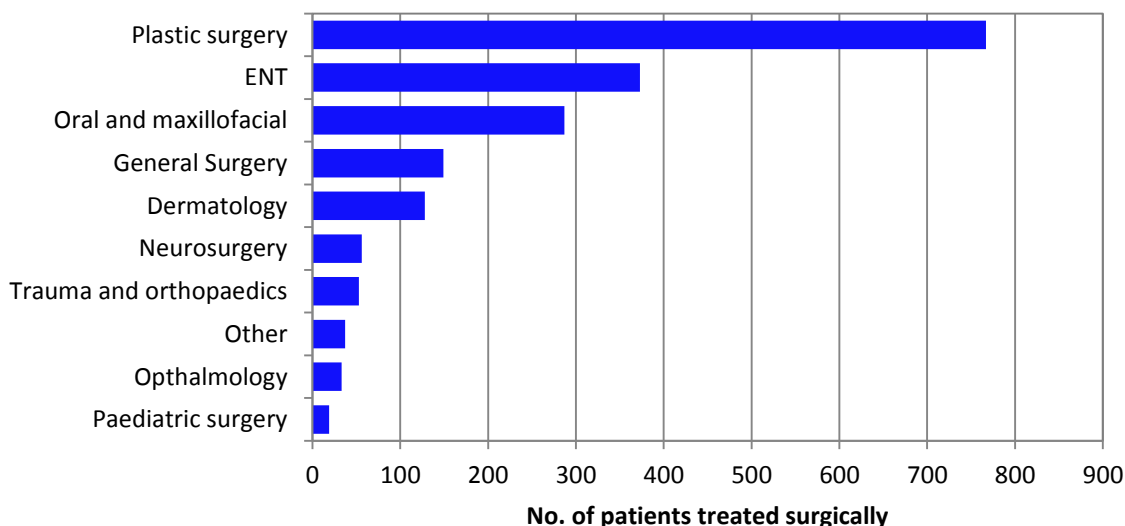


Figure 17: Number of head and neck soft tissue sarcomas treated surgically by each consultant specialty (England: 2000-2010)

Consultants specialising in plastic surgery oversaw the surgical management of 40% of patients with head and neck soft tissue sarcomas, with a further 20% overseen by ear, nose and throat (ENT) specialists and 15% by oral and maxillofacial surgeons (Figure 17). Table 3 shows the average time in days between diagnosis and first admission for surgical treatment, for the specialties of the MDTs within the hospital Trusts providing curative surgical procedures. Patients treated surgically within a hospital hosting a head and neck cancer MDT, on average, received treatment within three weeks of diagnosis. Those treated within a hospital trust hosting a skin cancer MDT received surgical treatment within 14 days of diagnosis.

Table 3: Average time between diagnosis and surgical treatment for each MDT hosted within hospital Trusts (England: 2000-2010)

MDTs within treatment centres	N	Avg days
HN/Sarcoma	466	37
BCNS/N	35	26
Childrens	23	23
HN	492	22
No specialism	89	18
Skin	337	14
Eye	5	9
Grand Total	1,447	25

Conclusions

This report describes incidence and survival rates for patients with bone and soft tissue sarcoma and highlights where patients were treated surgically between 2000 and 2010. The analyses demonstrates the impact that the Head and Neck Cancer IOG had on the referral of these patients into the larger hospital Trusts which host head and neck cancer MDTs. Within the data currently available it is not possible to establish whether patients with head and neck sarcoma were discussed by a head and neck cancer MDT or a sarcoma MDT, although this information should be collected by the English National Cancer Registration Service from April 2016 onwards.

Bone and soft tissue sarcomas of the head and neck region are a rare group of heterogeneous neoplasms. Around 38 bone sarcomas and 190 soft tissue sarcomas arising in the head and neck region are diagnosed annually. Age specific incidence rates increase with age. The incidence of head and neck soft tissue sarcomas is 60 per million in males aged 85 years and over, five times the rate observed in females of the same age. Rhabdomyosarcoma and angiosarcoma collectively account for 4% and 3% of sarcomas diagnosed² respectively, although they represent 13.6% and 12.5% of sarcomas arising in the head and neck region. These tumours are notorious for being highly aggressive with a high rate of tumour-related death¹⁰. Five-year relative survival rates are slightly higher for patients with bone sarcoma (73%) than for those with soft tissue sarcoma (64%).

Although rare, sarcomas of the head and neck region are an important group of tumours as it is often difficult to obtain wide surgical margins (due to the anatomical location) and they are thus prone to a high rate of recurrence⁴. It has also been shown that involved surgical margins are an important prognostic factor affecting long term outcomes for patients¹¹.

The results presented in this report demonstrate the high number of hospital Trusts involved in the surgical management of head and neck sarcomas, including bone sarcomas for which there are only five nationally commissioned specialist centres. However, following the release of the IOG for Patients with Head and Neck Sarcomas in 2004, the proportion of patients treated surgically in a hospital Trust hosting a head and neck cancer MDT increased significantly from 63% in 2000-2004 to 70% in 2005-2010. In the latter period, 139 different hospital Trusts oversaw the surgical management of at least one patient with a soft tissue sarcoma of the head and neck region. Further investigation of head and neck sarcoma treatment pathways is also essential in order to understand delays in hospital referrals for patients with head and neck sarcoma.

References

1. Ries, L. A. G., Kosary, C. L. and Hankey, B. F. (1999). SEER Cancer Statistics Review 1973-1996. National Cancer Institute, Bethesda, MD. Available online at: http://seer.cancer.gov/csr/1975_2000/.
2. National Cancer Intelligence Network (2012). Soft tissue sarcoma: Incidence and survival. Available online at: <http://www.ncin.org.uk/view?rid=2062> [accessed: 09/10/2013].
3. Dorfman, H. D. and Czerniak, B. (1995). Bone cancers. *Cancer*, 75 (S1), 2003-210.
4. Singh, R. P., Grimer, R. J., Bhujel, N., Carter, S. R., Tillman, R. M. and Abudu, A. (2008). Adult Head and Neck Soft Tissue Sarcomas: Treatment and Outcome. *Sarcoma*, 2008; 2008: 654987.
5. National Institute for Health and Clinical Excellence (2006). Guidance on Cancer Services: Improving Outcomes in Head and Neck Cancers: The Manual.
6. National Institute for Health and Clinical Excellence (2006). Guidance on Cancer Services: Improving Outcomes for People with Sarcoma.
7. Pollock, R. E. (2002). Atlas of Clinical Oncology. BC Decker Inc: London, page 127.
8. Pollock, R. E. (2002). Atlas of Clinical Oncology. BC Decker Inc: London, page 125.
9. Kraus DH, Sarenz NC, Gollamudi S, Heller G. Paediatric rhabdomyosarcoma of the head and neck. *Am J Surg* 1997; 174:556-60.
10. Fletcher D.M., Bridge, J.A., Hogendoorn, P.C.W. (2013) WHO Classification of Tumours of Soft Tissue and Bone, International Agency for Research on Cancer, Lyon, page 158
11. De Bree, R., van der Valk, P., Kuik, D. J. et al (2006) Prognostic factors in adult soft tissue sarcomas of the head and neck: a single-centre experience. *Oral Oncol*, 42(7); 703-709

Appendices

APPENDIX A - HEAD AND NECK ICD-10 CANCER SITE CODES

ICD10 group	ICD10 code	Description
Bones	C410	Malignant neoplasm of bones of skull and face
	C411	Malignant neoplasm of bone and articular cartilage of mandible
Skin	C440	Skin of lip
	C441	Skin of eyelid, including canthus
	C442	Skin of ear and external auricular canal
	C443	Skin of other and unspecified parts of face
	C444	Skin of scalp and neck
Connective tissue and nerves	C490	Connective and soft tissue of head, face and neck
	C470	Peripheral nerves of head, face and neck
Glands	C73	Malignant neoplasm of thyroid gland
	C751	Pituitary gland
	C753	Pineal gland
Undefined	C760	Malignant neoplasm of other and ill-defined sites: Head, face and neck
Eye	C69	Malignant neoplasm of eye and adnexa
Oral	C00	Malignant neoplasm of lip
	C01	Malignant neoplasm of base of tongue
	C02	Malignant neoplasm of other and unspecified parts of tongue
	C03	Malignant neoplasm of gum
	C04	Malignant neoplasm of floor of mouth
	C05	Malignant neoplasm of palate
	C06	Malignant neoplasm of other and unspecified parts of mouth
Head and neck	C07	Malignant neoplasm of parotid gland
	C08	Malignant neoplasm of other and unspecified major salivary glands
	C09	Malignant neoplasm of tonsil
	C10	Malignant neoplasm of oropharynx
	C11	Malignant neoplasm of nasopharynx
	C12	Malignant neoplasm of pirform sinus
	C13	Malignant neoplasm of hypopharynx
	C14	Malignant neoplasm of other and ill-defined sites in the lip, oral cavity and pharynx
Nasal, sinuses and throat	C30	Malignant neoplasm of nasal cavity and middle ear
	C31	Malignant neoplasm of accessory sinuses
	C32	Malignant neoplasm of larynx
	C33	Malignant neoplasm of trachea

APPENDIX B – SARCOMA ICD-03 MORPHOLOGY CODES

Morphology	Description
8710	Glomangiosarcoma: Glomoid sarcoma
8711	Glomus tumour (and variants), 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 epithelioid 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	Metastising leiomyosarcoma
8900	Rhabdomyosarcoma, NOS; rhabdosarcoma
8901	Pleomorphic rhabdomyosarcoma
8902	Mixed type rhabdomyosarcoma

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

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

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

Morphology	Description
9251	Giant cell tumour of soft parts, NOS
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 carcinomsarcoma
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 C – HEAD AND NECK CURATIVE SURGICAL TREATMENT CODES

OPCS4 Code	Surgery Description
A013	Partial lobectomy of brain
A021	Excision of lesion of tissue of frontal lobe of brain
A022	Excision of lesion of tissue of temporal lobe of brain
A023	Excision of lesion of tissue of parietal lobe of brain
A024	Excision of lesion of tissue of occipital lobe of brain
A025	Excision of lesion of tissue of cerebellum
A026	Excision of lesion of tissue of brain stem
A028	Excision of lesion of tissue of tissue of brain, other specified
A029	Excision of lesion of tissue of brain, unspecified
A178	Other specified therapeutic endoscopic operations on ventricle of brain
A293	Excision of lesion of trigeminal nerve (v)
A294	Excision of lesion of facial nerve (vii)
A298	Excision of lesion of specified cranial nerve NEC
A299	Unspecified excision of lesion of cranial nerve
A381	Extirpation of lesion of meninges of cortex of brain
A382	Extirpation of lesion of meninges of sphenoidal ridge of cranium
A383	Extirpation of lesion of meninges of subfrontal region of brain
A384	Extirpation of lesion of meninges of parasagittal region of brain
A388	Extirpation of lesion of meninges of brain, other specified
A389	Extirpation of lesion of meninges of brain, unspecified
A431	Extirpation of lesion of meninges of skull base
A432	Extirpation of lesion of meninges of skull clivus
A442	Extirpation of lesion of spinal cord NEC
A443	Excision of lesion of intramedullary spinal cord
A444	Excision of lesion of extradural spinal cord
A511	Extirpation of lesion of meninges of spinal cord
A571	Extirpation of lesion of spinal nerve root
A591	Total sacrifice of peripheral nerve
A598	Excision of peripheral nerve, other specified
A599	Excision of peripheral nerve, unspecified (includes Neurectomy nec)
A611	Excision of lesion of peripheral nerve
A619	Excision of lesion of peripheral nerve, unspecified
A753	Excision of lumbar sympathetic nerve
B012	Transphenoidal hypophysectomy
B014	Transcranial hypophysectomy
B041	Excision of lesion of pituitary gland, craniopharyngeal duct
B068	Excision of pineal gland, other specified
B081	Total thyroidectomy
B082	Subtotal thyroidectomy
B083	Hemithyroidectomy
B084	Lobectomy of thyroid gland NEC

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

OPCS4 Code	Surgery Description
B086	Partial thyroidectomy NEC
B088	Other specified excision of thyroid gland
B089	Thyroidectomy NEC, Unspecified
B121	Excision of lesion of thyroid gland
C011	Exenteration of orbit
C012	Enucleation of eye
C013	Evisceration of eye
C021	Excision of lesion of orbit
C028	Other specified extirpation of lesion of orbit
C101	Excision of lesion of eyebrow
C111	Excision of lesion of canthus
C121	Excision of lesion of eyelid NEC
C391	Excision of lesion of conjunctiva
C664	Laser photocoagulation of ciliary body
C821	Cauterisation of lesion of retina
D011	Total excision of external ear
D012	Partial excision of external ear
D018	Other specified excision of external ear
D021	Excision of lesion of external ear
D022	Destruction of lesion of external ear
D081	Extirpation of lesion of external auditory canal
D101	Radical mastoidectomy NEC
D103	Cortical mastoidectomy
D104	Simple mastoidectomy
D105	Excision of lesion of mastoid
D191	Excision of lesion of middle ear
E011	Total excision of nose
E018	Other specified excision of nose
E031	Submucous excision of septum of nose
E032	Excision of lesion of septum of nose
E042	Excision of turbinate of nose NEC
E043	Excision of lesion of turbinate of nose NEC
E081	Polypectomy of internal nose
E082	Extirpation of lesion of internal nose NEC
E091	Excision of lesion of external nose
E092	Destruction of lesion of external nose NEC
E132	Excision of lesion of maxillary antrum
E141	External frontoethmoidectomy
E142	Intranasal ethmoidectomy
E143	External ethmoidectomy
E144	Transantral ethmoidectomy
E171	Excision of nasal sinus NEC
E172	Excision of lesion of nasal sinus NEC
E191	Total pharyngectomy

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

OPCS4 Code	Surgery Description
E192	Partial pharyngectomy, Pharyngectomy NEC
E198	Other specified excision of pharynx
E201	Total Adenoidectomy, Adenoidectomy, Excision of Adenoid
E231	Open excision of lesion of pharynx
E232	Operations on pharyngeal pouch
E238	Other specified other open operations on pharynx
E241	Endoscopic extirpation of lesion of nasopharynx
E242	Endoscopic extirpation of lesion of pharynx NEC
E248	Other specified therapeutic endoscopic operations on pharynx
E291	Total laryngectomy
E292	Partial horizontal laryngectomy
E293	Partial vertical laryngectomy
E294	Partial laryngectomy NEC
E295	Laryngofissure and chordectomy of vocal chord
E296	Laryngectomy NEC
E301	Excision of lesion of larynx using thyrotomy as approach
E302	Excision of lesion of larynx using lateral pharyngotomy as approach
E309	Excision of lesion or larynx, Unspecified
E341	Microtherapeutic endoscopic extirpation of lesion of larynx using laser
E342	Microtherapeutic endoscopic resection of lesion of larynx NEC
E343	Microtherapeutic endoscopic destruction of lesion of larynx NEC
E348	Other specified microtherapeutic endoscopic operations on larynx
E352	Endoscopic resection of lesion of larynx
E353	Endoscopic destruction of lesion of larynx
E391	Open excision of lesion of trachea
E399	Unspecified partial excision of trachea
F019	Unspecified partial excision of lip
F021	Excision of lesion of lip
F201	Excision of gingiva, Gingivectomy NEC
F202	Excision of lesion of gingiva
F221	Total glossectomy
F222	Partial glossectomy, partial excision of tongue, wedge excision of tongue
F231	Excision of lesion of tongue
F232	Destruction of lesion of tongue
F281	Excision of lesion of palate
F288	Other specified extirpation of lesion of palate
F341	Bilateral dissection tonsillectomy
F344	Bilateral Tonsillectomy NEC, Bilateral excision of tonsil NEC
F348	Other specified excision of tonsil
F349	Tonsillectomy NEC
F381	Excision of lesion of floor of mouth
F382	Excision of lesion of mouth NEC
F384	Destruction of lesion of mouth NEC
F388	Other specified extirpation of lesion of other part of mouth

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

OPCS4 Code	Surgery Description
F389	Unspecified extirpation of lesion of other part of mouth
F441	Total excision of parotid gland
F442	Partial excision of parotid gland
F443	Excision of parotid gland NEC, Superficial parotidectomy
F444	Excision of submandibular gland
F448	Other specified excision of salivary gland
F449	Unspecified excision of salivary gland
F451	Excision of lesion of parotid gland
F452	Excision of lesion of submandibular gland
F453	Excision of lesion of sublingual gland
F454	Excision of lesion of salivary gland NEC
F459	Unspecified extirpation of lesion of salivary gland
F531	Open operations on parotid duct NEC
G038	Partial excision of oesophagus, Other specified
G041	Excision of lesion of oesophagus
G179	Endoscopic extirpation of lesion of oesophagus using rigid oesophagoscope, Unspecified
S048	Other specified other excision of skin
S049	Unspecified other excision of skin
S051	Microscopically controlled excision of lesion of skin of head or neck using fresh tissue technique
S053	Microscopically controlled excision of lesion of skin of head or neck using chemosurgical technique
S055	Microscopically controlled excision of lesion of skin of head or neck NEC
S058	Other specified microscopically controlled excision of lesion of skin
S059	Unspecified microscopically controlled excision of lesion of skin
S061	Marsupialisation of lesion of skin of head or neck
S063	Shave excision of lesion of skin of head or neck
S064	Shave excision of lesion of skin NEC
S065	Excision of lesion of skin of head or neck NEC
S066	Re-excision of skin margins of head or neck
S068	Repeat excision to clear margins, Other specified other excision of lesion of skin
S069	Unspecified other excision of lesion of skin
S081	Curettage and cauterisation of lesion of skin of head or neck
S082	Curettage and cauterisation of lesion of skin NEC
S083	Curettage of lesion of skin of head or neck NEC
S089	Unspecified curettage of lesion of skin
S091	Laser destruction of lesion of skin of head or neck
S101	Cauterisation of lesion of skin of head or neck NEC
S111	Cauterisation of lesion of skin NEC
T531	Excision of lesion of fascia
T652	Excision of lesion of tendon
T658	Other specified excision of tendon
T772	Wide excision of muscle
T773	Partial excision of muscle NEC
T779	Unspecified excision of muscle
T831	Destruction of lesion of muscle

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

OPCS4 Code	Surgery Description
T961	Excision of cystic hygroma
T962	Excision of lesion of soft tissue NEC
V051	Extirpation of lesion of cranium
V061	Medial maxillectomy
V068	Other specified excision of maxilla
V069	Maxillectomy, Unspecified excision of maxilla
V071	Extensive excision of bone of face
V072	Partial excision of bone of face NEC
V073	Excision of lesion of bone of face
V074	Excision of lesion of infratemporal fossa
V078	Other specified excision of bone of face
V079	Unspecified excision of bone of face
V141	Hemimandibulectomy
V142	Extensive excision of mandible NEC
V143	Partial excision of mandible, partial excision of jaw bone
V144	Excision of lesion of mandible
V148	Other specified excision of mandible
V149	Mandibulectomy nec, Unspecified excision of mandible
V221	Primary anterior decompression of cervical spinal cord and fusion of joint of cervical spine
V224	Primary anterior corpectomy of cervical spine with reconstruction HFQ
V228	Other specified primary decompression operations on cervical spine
V242	Primary decompression of thoracic spinal cord NEC
V292	Primary hemilaminectomy excision of cervical intervertebral disc
V294	Primary anterior excision of cervical intervertebral disc and interbody fusion of joint of cervical spine
V431	Excision of lesion of cervical vertebra
W085	Partial excision of bone NEC
W088	Other specified other excision of bone
W089	Unspecified other excision of bone
W091	Excision of lesion of bone
W341	Autograft of bone marrow
W348	Other specified graft of bone marrow
W761	Excision of ligament

APPENDIX D – SUPPORTING TABLES FOR FIGURES 1 - 17

Appendix D1 – supporting data for Figure 1

Cancer Site	Males		Females	
	Count	%	Count	Females
Bones	449	14%	344	21%
Connective tissue and nerves	1422	45%	593	36%
Skin	612	19%	220	13%
Nasal, sinuses and throat	251	8%	134	8%
Head & neck	152	5%	113	7%
Oral	107	3%	86	5%
Eye	85	3%	63	4%
Glands	33	1%	56	3%
Undefined	41	1%	35	2%

Appendix D2 – supporting data for Figure 2

Cancer Site	Males	Females	Total	Incidence ratios
Bones	449	344	793	1.31
Connective tissue and nerves	1,422	593	2,015	2.40
Skin	612	220	832	2.78
Nasal, sinuses and throat	251	134	385	1.87
Head & neck	152	113	265	1.35
Oral	107	86	193	1.24
Eye	85	63	148	1.35
Glands	33	56	89	0.59
Undefined	41	35	76	1.17
All sites	3,152	1,644	4,796	1.92

Appendix D3 – supporting data for Figure 3

Age group	Male to female incidence ratio	
	Bones	Soft tissue
0-9	1.15	1.28
10-19	1.32	1.06
20-29	1.31	1.44
30-39	1.46	1.22
40-49	1.08	1.67
50-59	1.82	1.61
60-69	1.95	2.54
70-79	0.95	3.17
80+	0.71	2.66

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

Appendix D4a – supporting data for Figure 4a

	Males			Females			Persons		
	ASR	LCI	UCI	ASR	LCI	UCI	ASR	LCI	UCI
1990	0.85	0.51	1.35	0.56	0.31	0.97	0.71	0.49	1.03
1991	0.58	0.31	1.00	0.65	0.35	1.10	0.61	0.42	0.92
1992	0.57	0.31	0.98	0.59	0.33	1.00	0.58	0.40	0.87
1993	0.40	0.18	0.78	0.38	0.18	0.74	0.39	0.24	0.65
1994	1.01	0.64	1.52	0.24	0.09	0.54	0.63	0.43	0.92
1995	0.58	0.32	0.97	0.37	0.17	0.72	0.47	0.31	0.74
1996	0.70	0.40	1.13	0.68	0.40	1.12	0.69	0.49	1.00
1997	0.63	0.36	1.04	0.49	0.27	0.87	0.56	0.38	0.84
1998	0.65	0.37	1.07	0.69	0.40	1.12	0.67	0.47	0.97
1999	0.63	0.35	1.05	0.62	0.35	1.03	0.62	0.43	0.92
2000	1.01	0.66	1.51	0.63	0.37	1.05	0.82	0.60	1.14
2001	0.84	0.52	1.31	0.78	0.47	1.24	0.81	0.59	1.14
2002	0.96	0.62	1.45	0.41	0.19	0.78	0.68	0.48	0.99
2003	0.57	0.32	0.97	0.54	0.29	0.93	0.55	0.38	0.83
2004	0.93	0.59	1.40	0.73	0.44	1.16	0.83	0.61	1.15
2005	1.26	0.86	1.80	0.60	0.34	1.01	0.93	0.69	1.27
2006	0.87	0.55	1.33	0.59	0.35	0.97	0.73	0.53	1.03
2007	1.28	0.88	1.81	0.88	0.56	1.34	1.08	0.82	1.43
2008	0.96	0.62	1.43	0.85	0.53	1.31	0.90	0.67	1.23
2009	1.10	0.74	1.59	0.64	0.38	1.05	0.87	0.65	1.19
2010	1.26	0.87	1.79	0.45	0.23	0.81	0.86	0.63	1.18

Appendix D4b – supporting data for Figure 4b

Gender	Age Group																	
	0004	0509	1014	1519	2024	2529	3034	3539	4044	4549	5054	5559	6064	6569	7074	7579	8084	85+
Males	0.3	0.2	0.5	0.5	0.8	0.6	0.9	0.8	0.7	0.8	1.1	1.5	1.4	1.6	1.8	1.4	2.2	1.1
Females	0.3	0.1	0.3	0.5	0.5	0.6	0.8	0.4	0.7	0.7	0.6	0.8	0.8	0.6	1.2	1.5	1.7	0.6
Persons	0.3	0.2	0.4	0.5	0.6	0.6	0.8	0.6	0.7	0.7	0.9	1.2	1.1	1.1	1.4	1.5	1.9	0.8

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Appendix D5a – supporting data for Figure 5a

	Males			Females			Persons		
	ASR	LCI	UCI	ASR	LCI	UCI	ASR	LCI	UCI
1990	4.34	3.55	5.28	1.57	1.11	2.18	2.96	2.50	3.51
1991	3.24	2.55	4.07	2.18	1.62	2.88	2.71	2.26	3.25
1992	3.54	2.82	4.39	2.12	1.60	2.78	2.83	2.39	3.37
1993	4.05	3.30	4.94	2.33	1.75	3.04	3.19	2.72	3.76
1994	3.49	2.79	4.33	2.08	1.56	2.74	2.79	2.35	3.32
1995	4.00	3.26	4.88	2.02	1.50	2.67	3.01	2.55	3.56
1996	4.29	3.52	5.20	1.87	1.38	2.49	3.08	2.62	3.63
1997	4.93	4.10	5.88	1.84	1.36	2.45	3.38	2.91	3.95
1998	5.42	4.55	6.41	1.51	1.08	2.09	3.47	2.98	4.04
1999	4.67	3.88	5.60	2.23	1.71	2.89	3.45	2.98	4.02
2000	4.37	3.59	5.27	2.69	2.08	3.43	3.53	3.04	4.11
2001	4.63	3.85	5.55	2.10	1.59	2.75	3.37	2.90	3.93
2002	4.79	4.00	5.72	1.77	1.29	2.39	3.28	2.82	3.84
2003	4.80	4.00	5.74	1.76	1.30	2.36	3.28	2.82	3.84
2004	5.53	4.68	6.50	2.20	1.67	2.88	3.86	3.37	4.46
2005	5.25	4.43	6.21	2.43	1.87	3.13	3.84	3.34	4.43
2006	5.25	4.45	6.18	1.98	1.50	2.60	3.62	3.15	4.17
2007	4.86	4.09	5.75	2.28	1.73	2.96	3.57	3.09	4.13
2008	5.75	4.91	6.71	2.28	1.74	2.95	4.01	3.51	4.60
2009	4.85	4.08	5.73	2.20	1.68	2.85	3.52	3.06	4.07
2010	5.08	4.30	5.96	2.48	1.92	3.17	3.78	3.30	4.34

Appendix D5b – supporting data for Figure 5b

Gender	Age Group																	
	0004	0509	1014	1519	2024	2529	3034	3539	4044	4549	5054	5559	6064	6569	7074	7579	8084	85+
Males	3.6	2.7	1.5	1.1	1.6	1.8	1.6	1.6	2.4	2.6	3.0	3.9	5.6	11.0	16.6	25.7	43.0	59.8
Females	2.9	2.3	1.5	1.0	1.3	1.1	1.1	1.5	1.6	1.4	2.2	2.1	2.6	3.4	4.1	6.0	7.7	10.7
Persons	3.2	2.5	1.5	1.0	1.4	1.5	1.4	1.6	2.0	2.0	2.6	3.0	4.1	7.0	9.8	14.2	20.7	24.6

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

Appendix D6 – supporting data for Figure 6

Cancer site	1993-98	1999-04	2005-10
Connective tissue and nerves	539	617	655
Skin	173	204	357
Nasal, sinuses and throat	103	113	124
Head and neck	75	82	81
Oral	46	51	68
Eye	36	49	43
Glands	30	27	23
Undefined	17	37	18
Bones	177	227	295
Grand Total	1,196	1,407	1,664

Appendix D7 – supporting data for Figure 7

Morphology	Connective tissue and nerves	Skin	Nasal, sinuses and throat	Head and neck	Oral	Eye	Glands	Undefined	Grand Total
Leiomyosarcoma	432	158	54	25	31	4	15	5	724
Angiosarcoma	319	128	17	11	10	3	7	7	502
RMS	164	9	100	98	55	106	2	11	545
Chondrosarcoma	15	0	67	0	0	1	0	3	86
Dermatofibrosarcoma	1	273	0	0	0	0	0	2	276
Fibrosarcoma	38	12	9	3	3	3	5	1	74
Liposarcoma	160	8	4	19	20	2	2	4	219
MFH	231	122	11	11	7	2	2	1	387
MPNST	171	4	3	3	2	1	0	2	186
Sarcoma, NOS	338	108	76	42	47	12	25	15	663
Other	146	10	44	53	18	14	31	25	341

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

Appendix D8 – supporting data for Figure 8

	Leiomyosarcoma	Angiosarcoma	Rhabdomyosarcoma	Chondrosarcoma	Dermato-fibrosarcoma	Fibrosarcoma	Liposarcoma	Malignant Fibrous Histiocytoma	Malignant peripheral nerve sheath tumour	Sarcoma, NOS	Other
0-9	-	-	305	0	-	5	-	-	7	11	26
10-19	-	-	87	-	8	-	0	0	7	21	28
20-29	17	-	53	-	25	-	-	-	22	36	40
30-39	10	7	31	5	53	10	17	6	19	32	36
40-49	35	9	25	9	53	5	40	5	22	41	44
50-59	52	29	16	20	37	9	44	13	22	49	48
60-69	104	85	10	16	28	11	54	61	32	110	37
70-79	247	162	11	21	35	13	40	127	20	157	51
80+	252	199	7	11	33	15	19	171	35	206	30
Total	722	502	545	86	276	74	219	387	186	663	340

Appendix D9a – supporting data for Figure 9

Year	n	Survival	LCI	UCI	Site
0		100%	100%	100%	STS
1	839	85%	82%	87%	STS
2	724	79%	75%	82%	STS
3	640	73%	69%	76%	STS
4	571	68%	64%	72%	STS
5	525	65%	61%	68%	STS
0		100%	100%	100%	bone
1	178	90%	85%	94%	bone
2	167	86%	80%	90%	bone
3	149	77%	70%	83%	bone
4	144	74%	66%	80%	bone
5	135	73%	65%	79%	bone

Head and neck sarcoma of the bone and soft tissue: Incidence, survival and surgical treatment

Appendix D9b – supporting data for Figure 9b

Year	n	Survival	LCI	UCI	Site
0		100%	100%	100%	STS- excluding skin
1	658	82%	79%	85%	STS- excluding skin
2	551	74%	70%	77%	STS- excluding skin
3	482	67%	63%	71%	STS- excluding skin
4	429	63%	58%	67%	STS- excluding skin
5	398	60%	56%	64%	STS- excluding skin
0		100%	100%	100%	bone
1	178	90%	85%	94%	bone
2	167	86%	80%	90%	bone
3	149	77%	70%	83%	bone
4	144	74%	66%	80%	bone
5	135	73%	65%	79%	bone

Appendix D10 – supporting data for Figure 10

Age Group	Surgery	No surgery	No HES	Grand Total
0-9	13	6		19
10-19	25	9	-	-
20-29	40	11	-	-
30-39	49	24	-	-
40-49	39	15	-	-
50-59	51	29	-	-
60-69	42	19	-	-
70-79	55	14	-	-
80+	15	17	-	-
Grand Total	329	144	18	491

Appendix D11 – supporting data for Figure 11

This contains 84 rows of data and is available on request

Appendix D12 – supporting data for Figure 12

Row Labels	2000-2004	2005-2010	Grand Total
No specialism	5	8	13
Childrens	7	8	15
BCNS\N	9	9	18
Skin	21	11	32
HN/Sarcoma	45	99	144
HN	68	96	164
Grand Total	155	231	386

Appendix D13 – supporting data for Figure 13

Specialism	Patients
Oral and maxillofacial	178
Neurosurgery	118
ENT	58
Other	18
Platic surgery	14
Grand Total	386

Appendix D14 – supporting data for Figure 14

Age group	Surgery	No surgery	No HES	Grand Total
0-9	62	113	-	-
10-19	49	45	-	-
20-29	68	34	5	107
30-39	81	30	16	127
40-49	110	49	15	174
50-59	139	54	7	200
60-69	211	90	22	323
70-79	338	140	49	527
80+	401	150	68	619
Grand Total	1,459	705	187	2,351

Appendix D15 – supporting data for Figure 15

This contains 160 rows of data and is available on request

Appendix D16 – supporting data for Figure 16

Specialist status	2000-2004	2005-2010	Grand Total
HN/Sarcoma	218	431	649
HN	241	400	641
Skin	171	242	413
No specialism	61	58	119
BCNS\N	23	20	43
Childrens	9	22	31
Eye		6	6
Grand Total	723	1,179	1,902

Appendix D17 – supporting data for Figure 17

Specialism	Patients
Plastic surgery	767
ENT	373
Oral and maxillofacial	287
General Surgery	149
Dermatology	128
Neurosurgery	56
Trauma and orthopaedics	53
Other	37
Ophthalmology	33
Paediatric surgery	19
Grand Total	1,902