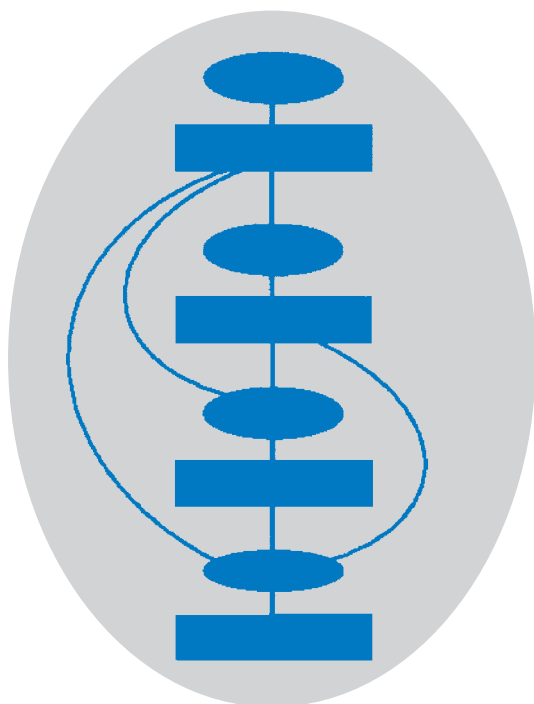


Guidance on Cancer Services

Improving Outcomes for People with Sarcoma

The Manual



March 2006

Developed by the National Collaborating Centre for Cancer

Improving Outcomes for People with Sarcoma

Cancer service guidance supports the implementation of *The NHS Cancer Plan* for England,¹ and the NHS Plan for Wales *Improving Health in Wales*.² The service guidance programme was initiated in 1995 to follow on from the Calman–Hine Report, *A Policy Framework for Commissioning Cancer Services*.³ The focus of the cancer service guidance is to guide the commissioning of services and is therefore different from clinical practice guidelines. Health services in England and Wales have organisational arrangements in place for securing improvements in cancer services and those responsible for their operation should take this guidance into account when planning, commissioning and organising services for cancer patients. The recommendations in the guidance concentrate on aspects of services that are likely to have significant impact on health outcomes. Both the objectives and resource implications of implementing the recommendations are considered. This guidance can be used to identify gaps in local provision and to check the appropriateness of existing services.

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This the latest in the series of Improving outcomes in cancer guidance documents deals with a group of relatively uncommon tumours. Because of their relative rarity there are particular challenges in ensuring that a clear diagnosis is made quickly and that patients get speedy access to the most skilled and appropriate advice and clinical care. We hope that the recommendations strike the appropriate balance between local and centralised specialist services, and will lead to changes in the provision of care that significantly improve the clinical outcomes and experience for these patients.

I am very grateful for the hard work and dedication of all the members of the Guidance Development Group who have worked so well and cheerfully together over the past 2 years, especially the patient representative members whose sensible reflections on their own and others' experiences have helped to shape the recommendations. I am also very grateful to the chair, Dr Joe Kearney, and the lead clinician, Mr Rob Grimer, whose skill, knowledge and dedication were invaluable in guiding the Group and completing the guidance.

Dr Fergus Macbeth

Introduction from the Chair of the Guidance Development Group

This guidance advises commissioners on how to improve the care of all patients with bone sarcomas and adults with soft tissue sarcomas. These tumours are relatively rare and can occur almost anywhere in the body, resulting in a wide variety of possible presentations. Although there are a number of important areas of care that are common to all these tumours, the management of patients with bone and soft tissue sarcomas involves quite distinct pathways of care. We have therefore addressed their needs separately. There are particular challenges in managing patients with these less common tumours, especially when some require very specialised surgical and other treatments, and this needs to be reflected in joint working both within and across cancer networks to achieve the important improvements in care that these patients require.

A number of soft tissue sarcomas will arise in children. These have traditionally been dealt with by paediatric oncologists and paediatric surgeons, adhering to strict protocols of management. Their management has also been covered in National Institute for Health and Clinical Excellence (NICE) guidance on 'Improving outcomes in children and young people with cancer',¹ and we have therefore excluded this group from this guidance.

Because of the rarity of these conditions the evidence base is not strong, but we believe that all the important and relevant evidence has been obtained and reviewed, and the appropriate conclusions drawn. The Guidance Development Group (GDG) consists of a wide range of experienced healthcare professionals and patients, the value of whose advice should not be underestimated. In addition, a survey undertaken by Sarcoma UK in 2004 helped to inform the GDG about the patient perspective on various aspects of the guidance. We believe that if this guidance is implemented, important and worthwhile changes will occur nationally in the management of these patients.

¹ National Institute for Health and Clinical Excellence (2005) Improving outcomes in children and young people with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

The format of the guidance is relatively simple. It starts with an epidemiological background and general clinical survey of sarcomas, and then outlines the current delivery of services in England and Wales. The main points presented include the rarity and diversity of these tumours, the increased incidence of bone sarcomas in younger patients, the differences between the pathways of care for patients with bone and for those with soft tissue sarcomas, and the large number of hospitals currently involved in delivering care to these patients.

The patient perspective follows. We have been as definitive in our advice in this part as we have been throughout the document. In particular we have made recommendations about what information should be supplied at different parts of the patient's pathway and who should be accountable for providing this.

The GDG spent a considerable time addressing the diagnostic pathway and believe that the recommendations in this area are those which will improve outcomes most significantly. The NICE 'Referral guidelines for suspected cancer' were our starting point.² Most patients with suspected sarcoma, as defined by these guidelines, will have a benign tumour, and the rapid assessment, identification and referral of those patients with malignant disease is the key to improving care and outcomes. We have described the diagnostic pathways that patients with bone or soft tissue sarcomas should follow, and expect that these recommendations will make a significant difference.

Expertise in radiology and histopathology is crucial for an accurate diagnosis and correct management. Simple recommendations such as the expert examination of a plain X-ray are important parts of the guidance. Although the provision of expert specialist pathologists is very important, such expertise is scarce. We have tried to deal with these issues without simply making facile recommendations about the provision of more consultant posts, and have suggested that expertise can be concentrated.

The recognition of the importance of a multidisciplinary team (MDT) approach to the care and management of all patients with cancer is now a 'given' in the NHS. The establishment of MDTs for all patients with sarcomas is recommended here. While most patients with bone sarcoma do currently have their care organised by an MDT, this is not the case for those with soft tissue sarcomas. The achievement of this recommendation would, with improvements in the diagnostic pathway, be of the greatest benefit in the care of these patients. The evidence for the optimal population base for such an MDT is not available, but our recommendations are logical and pragmatic. The existing teams throughout England and Wales should understand the basis of our decision and be able to work cooperatively to address this requirement.

² National Institute for Health and Clinical Excellence (2005) Referral guidelines for suspected cancer. *NICE clinical guideline* no. 27. Available from: www.nice.org.uk/CG027

We have proposed that the MDTs become not only responsible for the management of patients but also instrumental in establishing efficient and effective pathways of care from primary care to definitive treatment and follow-up. In essence the guidance recommends the establishment of managed sarcoma networks. We hope we have provided sufficient advice about this while still allowing some flexibility in the way in which healthcare professionals within cancer networks address the recommendations.

Radiotherapy and chemotherapy are important modes of treatment for these patients. Here the need for more local access has to be balanced against the advantages of concentrating these treatments in a few centres. We have managed this balance by recommending the most appropriate patient groups to the most appropriate place of treatment.

This diverse group of tumours can occur almost anywhere in the body. We have addressed all the main areas where MDTs from different disciplines will need to work closely together. We have supported this joint working without introducing a step that could seem bureaucratic, and at worst slow down the delivery of the best treatment for an individual patient. It should be recognised that for a minority of patients with sarcomas, there is a need for even more specialisation, and we have made this recommendation. As managed sarcoma care develops in England and Wales, further subspecialisation may well be established. We have paid particular attention to gastrointestinal stromal tumours (GIST). GIST has only recently been recognised as a distinct tumour type and even more recently has the role of novel agents, such as imatinib, been defined. Commissioners, we think, will appreciate its specific inclusion.

Support for patients with cancer from a wide range of professionals is fundamental to the delivery of high-quality care. Furthermore, as the need for quick and efficient steps in the pathway of care is recognised and underpinned by government targets, the support both to the individual patient with a key worker and to the MDT with administrative input is vital. For patients requiring limb amputation, the recommendations about the provision of high-quality prostheses and rehabilitation is another important aspect of this guidance. We have included advice on follow-up and on supportive and palliative care, where the recommendations complement the NICE guidance on 'Improving supportive and palliative care for adults with cancer'.³ We have tried to avoid too much duplication.

³ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

The final chapter – ‘Improving knowledge’ – is important in ensuring that the recommendations, when implemented, result in a sustained development of the delivery of care. The provision of a range of information and audits will demonstrate that this guidance was only the beginning of the process of developing care. Improving knowledge, at all levels, is fundamental to providing healthcare professionals and commissioners with a clear picture of the quality of care that they are delivering and of the need for continued improvements. Without this knowledge, change will be hampered; with it, we have the opportunity to demonstrate the delivery of high-quality care based on a sound and constantly improving evidence base. Without such information commissioners are unlikely to provide the necessary increases in resources that this patient group requires.

The economic analysis tries to estimate the cost impact of implementing the key recommendations of this guidance. These give some indicative costs which commissioners can use to carry out work to determine what the effects would be locally. In addition we are aware that the HRGs currently in use for major surgery on patients with soft tissue sarcoma may significantly underestimate the true costs of the procedure. The commissioners need to take this into account and there is an urgent need for this problem to be properly investigated at a national level.

Key recommendations

- All patients with a confirmed diagnosis of bone or soft tissue sarcoma (except children with certain soft tissue sarcomas) should have their care supervised by or in conjunction with a sarcoma multidisciplinary team (MDT).
- Cancer networks should arrange diagnostic services for the investigation of patients with suspected soft tissue sarcomas (as defined by the National Institute for Health and Clinical Excellence (NICE) 'Referral guidelines for suspected cancer') at designated diagnostic clinics. All patients with a probable bone sarcoma (usually following X-ray examination) should be referred directly to a bone tumour treatment centre (see Chapter 6) for diagnosis and management.
- All patients with a provisional histological and/or radiological diagnosis of bone or soft tissue sarcoma should have their diagnosis reviewed by a specialist sarcoma pathologist and/or radiologist who are part of a sarcoma MDT. Commissioners should fund a formal system for second opinions and review of difficult cases, and molecular pathology and cytogenetic facilities.
- A soft tissue sarcoma MDT should meet minimum criteria (as defined in Chapter 5) and manage the care of at least 100 new patients with soft tissue sarcoma per year. If a sarcoma MDT manages the care of patients with both bone and soft tissue sarcoma, it needs to manage the care of at least 50 new patients with bone sarcoma per year and at least 100 new patients with soft tissue sarcoma per year.
- All patients who are managed by a sarcoma MDT should be allocated a key worker (see Chapter 8).
- Patients should undergo definitive resection of their sarcoma by a surgeon who is a member of a sarcoma MDT or by a surgeon with tumour site-specific or age-appropriate skills, in consultation with the sarcoma MDT.

- Chemotherapy and radiotherapy are important components of the treatment of some patients and should be carried out at designated centres by appropriate specialists as recommended by a sarcoma MDT.
- Patients should be informed about relevant clinical trials and supported to enter them.
- All sarcoma MDTs should participate in national audit, data collection and training.
- Patients with functional disabilities as a consequence of their sarcoma should have timely access to appropriate support and rehabilitation services.
- The National Specialist Commissioning Advisory Group should consider commissioning designated centres for the management of retroperitoneal and pelvic soft tissue sarcomas.
- This guidance should be implemented by primary care trusts (PCTs)/local health boards (LHBs) working collaboratively through their specialist commissioning groups, in close consultation with cancer networks. A National Implementation Group should be considered for both England and Wales.

Background

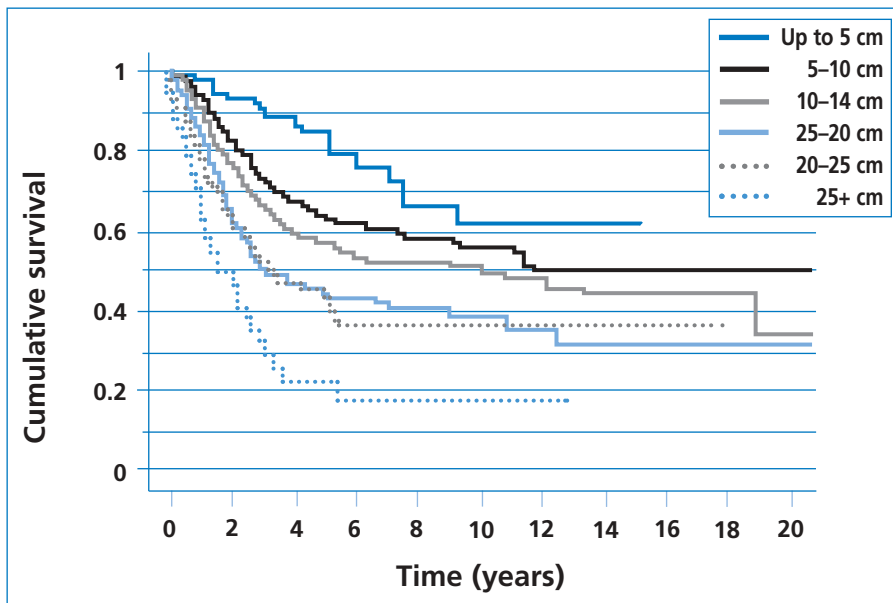
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Sarcomas are a rare and diverse group of cancers thought to have a common embryological origin. They arise from cells that make up the connective tissue structure, including bone, cartilage, muscle, blood vessels, nerves and fat. Sarcomas can be broadly divided into those of bone and those of soft tissue (STS). STS are ranked the 23rd most common cancer type and bone tumours the 27th most common type. If combined, sarcomas would still only be the 21st most common cancer type. [1]

During a working lifetime a general practitioner (GP) with a list size of 2000 patients may see many hundreds of benign tumours, but can only expect to see one or two patients with bone or soft tissue sarcomas. [2] Even within secondary care the majority of patients seen with soft tissue tumours are likely to have a benign lesion, so identifying the small number of patients with sarcoma generates a considerable diagnostic workload for clinicians.

Delays in diagnosis for both bone and STS are common. The median size on presentation for both bone and STS is 10 cm (Royal Orthopaedic Hospital: unpublished data 2005) – earlier diagnosis would undoubtedly lead to improved outcomes in terms both of survival and of less-damaging surgery being required (Figure 1). Many STS are discovered incidentally following excision of a lump, with no prior suspicion that it could be a sarcoma. Very often this initial excision is inadequate and further treatment is required.

Figure 1. Survival of all sarcomas without metastases at diagnosis, split by size category at diagnosis



Source: Royal Orthopaedic Hospital: unpublished data 2005.

Incidence of sarcomas in England and Wales

The coding system used for cancers is the International Classification of Diseases, currently in its 10th edition (ICD-10). [3] The ICD-10 classifies cancers to specific body sites with codes for tumours of bone and for tumours of connective and soft tissue. (Childhood cancers are also coded using the International Classification of Childhood Cancer (ICCC).) The ICD-10 classification was used because it is available for both adults and children.

- C40 malignant neoplasm of bone and articular cartilage of limbs
- C41 malignant neoplasm of bone and articular cartilage of other and unspecified sites
- C49 malignant neoplasm of connective and soft tissue

Data from ICD-10

Ideally all cancers, of whatever type, are recorded by the cancer registries in England and Wales and over 90% ascertainment is currently reported. [4] For the 5-year period between 1996 and 2000 there was an average of 427 primary bone tumours registered each year in England and Wales under codes C40 and C41 (Table 1).

Table 1. Registrations, crude incidence, deaths and mortality for primary bone cancers

Site of tumour	Average annual number of registrations England and Wales 1996–2000	Incidence (crude rate per million)	Number of deaths (2002)	Mortality (crude rate per million)
C40 and C41	427	8.2	242	4.6

Data from Office for National Statistics (ONS) and Welsh Cancer Intelligence and Surveillance Unit.

For a similar period, an average of 1094 connective and soft tissue tumours were registered each year in England and Wales (Table 2) under C49. As these tumours can arise from connective tissue in sites all over the body, they may be coded to the sites where they occur rather than to the connective tissue (C49) category. This potentially leads to an underestimate of the number of soft tissue sarcomas if using cancer registry data.

Table 2. Registrations, crude incidence, deaths and mortality for connective and soft tissue cancers

Site of tumour	Average annual number of registrations England and Wales 1996–2000	Incidence (crude rate per million)	Number of deaths (2002)	Mortality (crude rate per million)
C49	1094	21.13	589	11.30

Data from Office for National Statistics (ONS) and Welsh Cancer Intelligence and Surveillance Unit.

Five-year data from the East Anglia Cancer Registry were reviewed for this guidance and it was found that only 53% of soft tissue sarcomas were coded to ICD-10 C49. Other sites included the uterus, gastrointestinal tract and retroperitoneum.

If coding practices are similar throughout England and Wales then approximately 2000 soft tissue sarcomas, in all sites, might be expected each year.

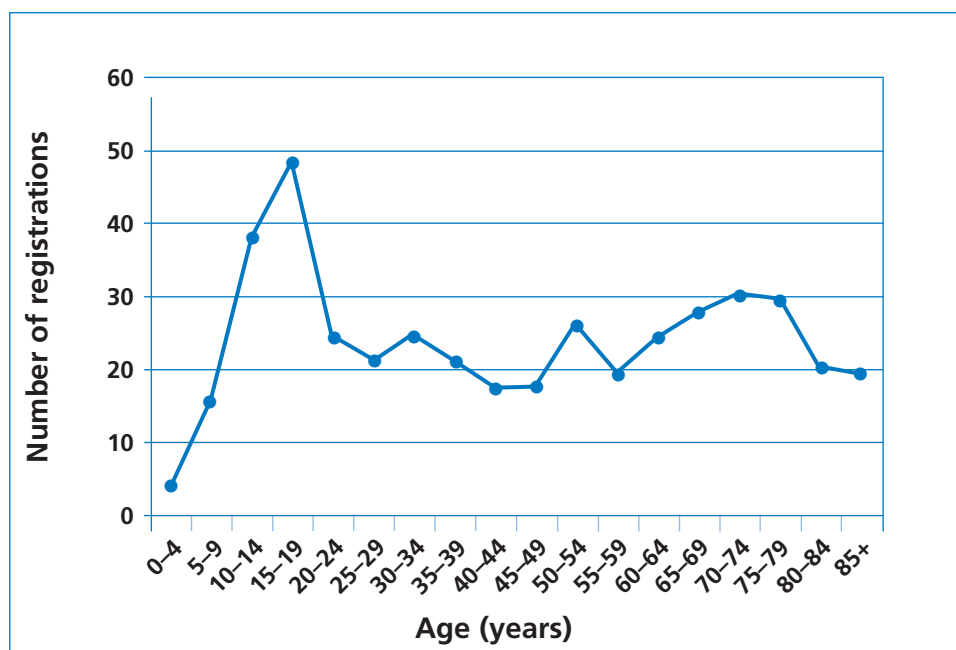
It is important to note here that the historical cancer registry data do not take into account the recent advances in the classification of gastrointestinal stromal tumours (GIST). These are a form of soft tissue tumour of particular interest because a targeted therapy (imatinib) has been developed. It has only recently been possible to classify GIST using immunohistochemistry, and data on incidence are not yet available. Estimates of incidence vary widely, from 4 to 40 cases per million population, although recent data from Sweden suggest the incidence is in the region of 15 per million per year. [5]

Bone sarcomas

Bone sarcomas are estimated to account for 0.2% of all malignant tumours, [6] but represent 4% of all malignancy in children aged up to 14 years. [7] The symptoms can vary, but the most common are pain and swelling or tenderness in the affected area. The cancer can cause weakness of the bone leading to a fracture. The presence of these symptoms should lead to referral for an X-ray, which in turn may lead to the suspicion of bone malignancy.

The age-specific frequencies of primary bone sarcomas are bimodal – the first peak occurring during the second decade of life, associated with the growth spurt, and the second occurring in patients older than 60 years. They are more common in males than in females. [7] Figure 2 shows the age-specific incidence for primary bone cancers.

Figure 2. Average annual number of registrations of primary cancers of bone and cartilage (1996–2000), England and Wales, by 5-year age bands



Data from Office for National Statistics (ONS) and Welsh Cancer Intelligence and Surveillance Unit.

The following are the most common histological types of malignant bone tumours:

- a) **osteosarcoma:** the most common primary malignant bone tumour. It occurs predominantly in patients younger than 20 years, in whom 80% of tumours occur in long bones of the extremities. [7] In the older age group osteosarcomas may arise secondary to radiation or Paget's disease
- b) **chondrosarcoma:** the incidence of this type of malignant bone tumour increases gradually with age. More than 50% of these tumours occur in long bones of the extremities. [8] They may also occur in the pelvis and ribs
- c) **Ewing's sarcoma:** the major peak for age-specific incidence occurs in the second decade of life with a rapid decrease after the age of 20 years. [7] These tumours typically arise in the axial skeleton (pelvis, scapula, rib) or in the diaphysis (main or mid section) of long bones
- d) **spindle cell sarcomas:** these are a variety of other rare sarcomas of bone, for example fibrosarcoma, malignant fibrous histiocytoma and leiomyosarcoma, which behave just like osteosarcoma but typically arise in an older population.

Risk factors for bone sarcomas

Although the majority of patients do not have any apparent risk factors there are a number of pre-existing conditions and exposures that have been associated with an increased risk of bone cancer.

Genetic conditions

Individuals with certain rare inherited cancer syndromes where cell regulatory genes are altered have an increased risk of developing osteosarcoma, for example in Li–Fraumeni syndrome (mutation of p53).

Precursor conditions

There are several precursor conditions that are associated with an increased risk of bone cancer. For example, Paget's disease of bone, a benign condition mostly affecting people older than 50 years, causes formation of abnormal bone tissue. Bone sarcomas (usually osteosarcoma) develop in about 5–10% of patients with severe Paget's disease. [9]

Benign bone tumours such as osteochondromas and enchondromas also have a slight risk of developing into a chondrosarcoma. Patients with multiple lesions, as found in hereditary multiple exostoses (HME), Ollier's disease or Maffucci's disease, are also at an increased risk. [10]

Radiation

Bone exposure to ionising doses of radiation increases the risk of developing bone cancer (for example, radiation therapy to treat another cancer). Treatment at a younger age and/or being treated with higher doses of radiation (usually over 60 Gy) increase the risk of developing bone cancer (usually osteosarcoma). [11]

Treatment of bone sarcomas

The main way of treating bone sarcomas is a combination of surgery and chemotherapy. Modern surgical treatment aims to achieve a complete removal of the primary tumour while at the same time preserving the limb and limb function (or other body part) wherever possible. Surgical treatment is often disabling, even when amputation has not been performed, and patients require rehabilitation, including physiotherapy and occupational therapy, to recover optimal personal and social functioning, including return to work. Some patients require lifelong provision of orthotic and/or prosthetic appliances.

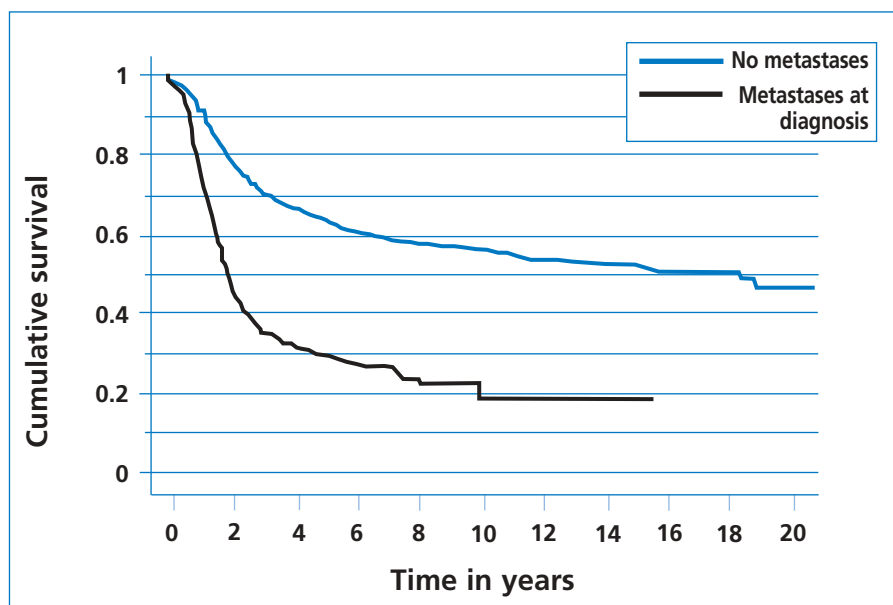
Chemotherapy regimens are used for bone sarcoma and are among the most complex in adult oncology practice. Adjuvant chemotherapy (given both preoperatively and postoperatively) contributes significantly to long-term survival for patients with Ewing's sarcoma and osteosarcomas. [12–16]

Radiotherapy is a key part of curative treatment for some patients with Ewing's sarcoma and is a valuable part of palliative therapy for other patients with bone sarcoma. Radiotherapy is typically delivered by fractionation of the total dose over 4 to 6 weeks, with daily attendances for treatment.

Prognosis and survival of bone sarcomas

Survival for patients with bone tumours has improved substantially over the last 30 years. A national study looking at the survival of adults aged 15 years or older with bone cancer of all types found 5-year relative survival rates increased from 29% in 1971–75 to 51% in 1986–90. [17] The most important prognostic factors include the presence of detectable metastases at diagnosis (Figure 3), tumour volume, increasing age and response to chemotherapy.

Figure 3. Survival of all patients with bone sarcomas, split by whether they have metastases or not at diagnosis



Source: Royal Orthopaedic Hospital: unpublished data 2005.

The European cancer registry-based study on survival and care of cancer patients aims to describe survival differences between European populations and the reasons for them. [18] For adults diagnosed between 1990 and 1994 the survival rates for primary bone tumours in England and Wales were not significantly different from the average for Europe (Table 3). (It should be noted that small numbers are involved and there are potential differences in the quality of data available in different countries.)

Table 3. Five-year survival for primary bone cancer in adults diagnosed between 1990 and 1994

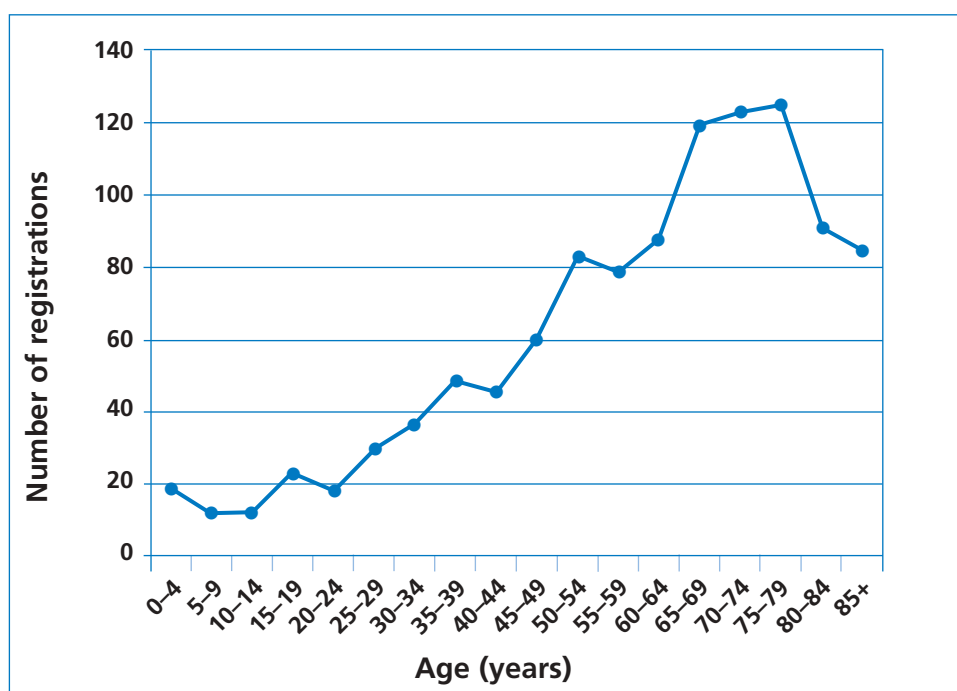
	Age-standardised relative survival (%) and 95% CI	
	Men	Women
England and Wales	51.4 (46.9–56.3)	54.8 (49.8–60.3)
Europe (average)	53.0 (48.0–58.4)	56.3 (51.5–61.6)

CI: confidence interval

Data from EUROCARE-3

Soft tissue sarcomas (STS) account for about 1% of all malignant tumours. [19] Benign soft tissue tumours outnumber malignant by at least a factor of 100. STS can occur anywhere that connective tissue is present and the signs and symptoms vary greatly depending on the anatomic site, as do the treatment options and prognosis. Soft tissue sarcomas increase in frequency with age (Figure 4).

Figure 4. Average annual number of registrations of connective and soft tissue cancers (C49), (1996–2000), by 5-year age bands, England and Wales



Data from Office for National Statistics (ONS) and Welsh Cancer Intelligence and Surveillance Unit.

The following are some common anatomical sites where STS can occur:

- a) **extremity and superficial trunk:** the majority of patients with STS of the extremities and superficial trunk present with a mass which is usually painless. It can be difficult to differentiate a benign from a malignant mass, but urgent referral guidelines have been produced by NICE to identify patients more likely to have a malignant tumour [2]

Features suggestive of malignancy in a lump include:

- lump >5 cm
- lump increasing in size
- lump deep to the fascia
- pain

NICE 'Referral guidelines for suspected cancer' (www.nice.org.uk/CG027)[2]

- b) **retroperitoneum:** most patients present with an abdominal mass, with half reporting pain at presentation. Because of the space available in the retroperitoneum, these tumours may often grow to a substantial size before presenting, and the overall prognosis is worse for people with retroperitoneal tumours than for those with extremity sarcomas
- c) **viscera:** sarcomas of the viscera present with signs and symptoms particular to the organ of origin. For example, GIST, which occur primarily in the middle-aged and older population, present with upper abdominal pain in 40–50% of cases. [20] Melaena, haematemesis or palpable tumour may also be presenting features. Sarcomas of the uterus often present with painless vaginal bleeding as occurs with other uterine malignancies
- d) **head and neck:** sarcomas can arise from bone, cartilage or the soft tissues of the head and neck. The majority occur in adults, but in children 40% of soft tissue sarcomas that occur arise in the head and neck region. [21] They can present as a lump, with problems relating to compression of the surrounding anatomy such as the orbit or pharynx. Surgery and radiotherapy are difficult because of the proximity of important anatomy in this area.

Risk factors of STS

As with bone sarcomas, in most cases of soft tissue sarcoma it is not possible to identify a specific aetiological agent. A number of genetic conditions including Li–Fraumeni syndrome, hereditary retinoblastoma, neurofibromatosis and familial adenomatous polyposis (Gardner's syndrome) carry an increased risk for soft tissue sarcoma.

Lymphoedema is associated with lymphangiosarcoma, most often after radical lymphadenectomy, but also in primary lymphoedema.

Treatment of STS

The treatment of STS is largely surgical – excising the tumour with an adequate margin of normal tissue around it. For patients with large and high-grade tumours, radiotherapy will also usually be used.

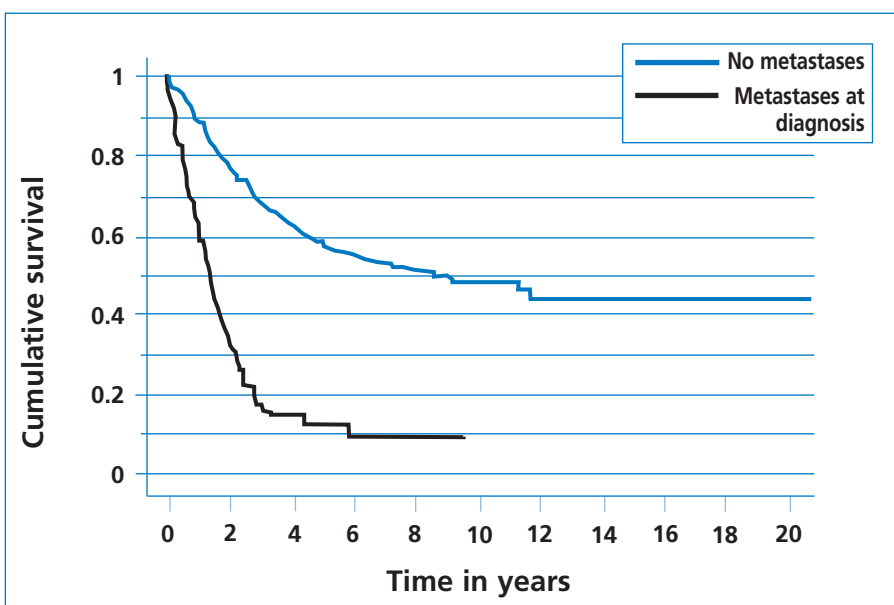
Chemotherapy is principally used for treating specific STS (soft tissue Ewing’s tumour, rhabdomyosarcomas and STS in children), but may be used in the treatment of large high-grade tumours to improve local control, having only a small unproven benefit on overall survival. [22]

Prognosis and survival of STS

Five-year survival is between 50% and 60% for soft tissue sarcomas as a group; [23] however there is wide variation depending on anatomical site and histological features of the tumours (including GIST). Data from EUROCORE-3 show 5-year survival for tumours coded to ‘soft tissue’ in England is not significantly different from that in Europe as a whole. [5]

The prognosis for patients with limb and trunk STS is based on five factors: the patient’s age, the presence of metastases at the time of presentation (Figure 5), the size of the tumour, its depth and its histopathological grade. Tumours of grade 1 are treated as low grade, and those of grades 2 and 3 are treated as high grade. In a large series, the proportion of grade 1, 2 and 3 were 16%, 28% and 56% respectively (Royal Orthopaedic Hospital: unpublished data 2005). Patients with retroperitoneal sarcomas have a poorer prognosis, largely because these tumours present so late. [24]

Figure 5. Survival of patients with extremity soft tissue sarcomas, split by whether they have metastases or not at diagnosis



Source: Royal Orthopaedic Hospital: unpublished data 2005.

Current services for sarcomas

People with bone sarcomas

The diagnosis and surgical treatment of primary bone tumours are very complex, and the two supraregional bone tumour treatment centres, set up by the National Specialist Commissioning Advisory Group (NSCAG) in 1984, play a central role in their management. One of the two centres is at the Royal Orthopaedic Hospital in Birmingham and the second centre is in London, split between University College Hospital and the Royal National Orthopaedic Hospital at Stanmore.

The original remit of these supraregional centres was to provide 'investigation and treatment for patients who may have primary malignant bone tumours requiring endoprosthetic replacement'. It is recognised that the definition of the supraregional service is now redundant because of the many different ways in which primary bone tumours can be managed apart from by endoprosthetic replacement. In 2005 NSCAG therefore widened the definition and will commission services for 'the diagnosis and treatment of primary malignant bone tumours'. Other units including Newcastle, Oxford, Bristol and Oswestry are now contracted to supply this service.

A survey of cancer networks carried out for this guidance found that referral pathways for patients with potential bone tumours are well established but diverse. However, they are not formalised, and may be influenced by professional relationships that have developed over time. Radiotherapy and chemotherapy for these patients is provided more widely. An analysis of 5 years of hospital episode statistics (HES for England and patient episode database, Wales (PEDW) for Wales) (1997/98 to 2001/02) identified 33 trusts that had more than 50 episodes coded to medical and/or clinical oncology for the treatment of primary bone tumours.

Most primary malignant bone tumours occur in adolescents or children, and these patients will receive their non-surgical treatment at a principal treatment centre for children and young people. They will however require complex surgery carried out at an NSCAG centre.

People with soft tissue sarcomas

The management of patients with STS is not designated by NSCAG and they are treated by a range of clinicians. In many cases there is no clear pathway for patients with suspected sarcoma. Many patients with STS (possibly half the total) are still treated in district hospitals by non-specialists. Delays in diagnosis are frequent, despite guidelines about early referral for possible malignancy.

A review of 5 years of hospital episode statistics (HES) and PEDW data carried out for this study identified 65 trusts in England and Wales with more than 50 hospital episodes over the period coded to orthopaedics, general and plastic surgery for sarcoma, and 189 trusts showing at least some activity. Also in this survey a total of 18 trusts reported that they performed planned surgery on soft tissue sarcomas and had a multidisciplinary team (MDT) in place. We are aware of at least another six trusts with an interest in sarcoma care. The Royal Marsden and Royal Orthopaedic Hospital reported the most activity, reflecting the known expertise in these centres. Responders to the survey also reported that the care of people with gynaecological sarcomas and sarcomas of the gastrointestinal tract is often managed by those specialty multidisciplinary teams.

The review of HES and PEDW data found 120 trusts providing non-surgical oncology treatment (radiotherapy and chemotherapy) to patients with soft tissue sarcoma.

Peer review

Until now there has been no national system of peer review of sarcoma treatment centres. There have been locally commissioned reviews, but these have been largely ‘generic’ with different standards across the country. What constitutes a sarcoma treatment centre in terms of standards, number of patients treated and staffing has not been consistently defined.

NSCAG does review the supraregional bone tumour treatment centres annually, and there is an annual combined audit meeting between the two original designated centres.

Training

There is no established system of training or accreditation for sarcoma surgery, or indeed for the other specialties involved. Most of the current consultants will have received training in this country or abroad at existing treatment centres.

Patient support

It is well recognised that patients require special support when affected by a malignant tumour. Because of the rarity of sarcomas, most patients and non-specialist clinicians will have no background knowledge of the condition. This can lead to a sense of frustration and isolation for the patient. The problem is not helped by fragmented information and few reliable information sources. There are also specific long-term healthcare support issues that must be addressed. Sarcoma surgery is frequently disabling or disfiguring, and although fewer patients face amputation than in the past they require lifetime access to support services with specific expertise. Some centres have established key workers, usually clinical nurse specialists, and patient support groups.

Key points

- These are rare tumours. There are approximately 400 patients diagnosed with bone sarcomas and 2000 with soft tissue sarcomas per year in England and Wales.
- The diagnosis is often not suspected before biopsy or excision.
- The diagnostic pathway is better described for bone tumours than for soft tissue sarcomas.
- Significant numbers of patients, especially those with STS, are probably not managed by a multidisciplinary team.
- The peak incidence for bone tumours occurs in children and young people.

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Treatment and care need to take into account patients' individual needs and preferences. People with sarcoma should have the opportunity to make informed decisions about their care and treatment.

Sarcomas are rare, and non-specialist doctors may have little or no experience of diagnosing or treating them. A patient may be reassured during the diagnostic process that the problem they are suffering from is not life-threatening, but this can change when the diagnosis is finally explained to them. This may sometimes even occur after they have had surgery. It can therefore be a shock when the diagnosis of sarcoma is finally made. There is also a shortage of good information to help the patient and their family begin to understand the disease, its treatment and the prospects they now face. Therefore there needs to be a clear focus on access to relevant, high-quality and timely information and emotional support from family and health professionals. Definitive guidance on all aspects of information and support is given in the NICE guidance on 'Improving supportive and palliative care for adults with cancer'.⁴

The variety of sarcomas and the range of anatomical sites at which they are found mean that each patient has very specific information needs at each stage of their pathway of care through diagnosis, treatment, follow-up and discharge.

A key worker (see Chapter 8) is valuable to this support, especially in answering questions about treatment and providing help at a time and place of stress. In addition the value of support from self-help groups is recognised in many cancers, and the anecdotal experience of the few sarcoma-specific self-help groups in the UK reflects this.

Because of the rarity of these tumours and the relatively small number of specialised treatment centres, many patients may have to travel long distances for their management. On balance, most patients wish to receive the best possible treatment that is available to them and will cope with the travel issues as a secondary pressure if necessary.

⁴ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

Information

The ability of patients to take in and remember sometimes complex information at a time of great stress is limited. This calls for an approach to information-giving which allows different 'techniques' to be used, such as:

- face-to-face with a specialist nurse or other healthcare professional
- leaflets addressing the patient's specific situation
- leaflets offering generic treatment information (for example, about chemotherapy)
- audio, video or CD resources
- the Internet
- telephone helpline with access to a specialist nurse or other healthcare professional.

It should not be assumed that patients have access to all information methods, or that all patients want all levels of information. Other than face to face, the main means of providing information will be a specific printed leaflet, supported by a healthcare professional who can guide the patient through the information, and who is able to address immediate questions, whether in person or by telephone. Such written information may be duplicated on a website or recorded onto audiotape, and may need to be translated into languages other than English, or made available in large-print versions.

Information can be considered as a number of 'layers' that can be accessed as required to provide the most appropriate information at that time, either by a healthcare professional or by the patient according to their own needs and wishes. At the simplest layer the information will be a very basic description, and at the most complex there may well be extended references to other resources, including the Internet. All layers can be supported by frequently asked questions (with appropriate answers) including questions that the patient can ask their own doctor.

The Internet presents a range of problems with regard to information quality. A patient using the Internet will inevitably come across sites that have negative messages, give a biased view, or are inaccurate. There is no easy answer to this problem. A short list of recommended sites (including why they are recommended and by whom) will go some way towards providing guidance.

Support

Patients may benefit from different kinds of support at different times during the course of their illness. Carers, especially close family, may also need psychological and social support.

Accepting support is a matter of choice, and patients should be able to choose for themselves. This may present issues when dealing with patients from ethnic or cultural groups where decisions may be traditionally taken within the family on behalf of a patient.

Choice and decision-making

There is a risk of providing too much information during the period following diagnosis, when choices have to be made at a time of great anxiety within deadlines determined by treatment resource availability. Patient decisions in such circumstances may be made on the basis of feelings, beliefs or values, which may or may not be disclosed to healthcare professionals. Crucial treatment decisions may be influenced by the distance from home to the treatment centre, and the means of transport available. There will be many issues that the patient must consider, and so support which can help them establish their personal priorities may be necessary. For example, the patient's decision may be influenced by whether the treatment is curative or palliative, or by something as simple as the provision of a reserved car-parking space.

The prospect of participating in a clinical trial can be an additional burden for some patients but will be welcomed by others. As many sarcoma trials are for drug treatments, it is important that the choices available are made clear to patients who are eligible for a trial.

GP information

GPs also need a reliable source of information. This would cover a number of areas such as raising awareness of sarcoma; providing information 'now you have a patient with sarcoma on your list'; actions with regard to lymphoedema, provision of nutritional advice etc; support for patients having problems with endoprosthetic implants; addressing specific issues relating to GIST and imatinib. This source of information could also be referred to in communication between treatment centres and the patients' GPs.

A. Recommendations

Diagnosis

A diagnosis or other significant news should be communicated by a senior doctor or specialist nurse who has enhanced skills (as defined in Chapters 3 and 4 of the NICE guidance on 'Improving supportive and palliative care for adults with cancer'⁵). Communication should be face to face unless there is specific agreement with the patient about receiving confirmation of a preliminary diagnosis by telephone or in writing.

All patients with a suspected or confirmed diagnosis of sarcoma should be allocated a key worker (see Chapters 5 and 8).

Patients should be offered a permanent written and/or audio record of their diagnosis and of any important points relating to the consultation. Their key worker and their contact points should be identified in writing and this information should also be supplied to their GP.

Information

Commissioners and provider organisations should ensure that at every diagnostic clinic/sarcoma treatment centre information is available that:

- is specific to that centre
- describes the tests/treatments it provides
- describes the individual patient's diagnosis or disease stage
- is age-appropriate (see the NICE guidance on 'Improving outcomes in children and young people with cancer'⁶).

Information should be provided in a variety of formats (for example, print/audio) and supported by information about access to online resources. Information should be written in language to which patients can directly relate. They should have as much information as they want, in a format that they can understand.

⁵ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

⁶ National Institute for Health and Clinical Excellence (2005) Improving outcomes in children and young people with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

All information should be developed and reviewed with the involvement of patients with sarcoma.

Table 4 maps the scope of the information which should be made available to patients at each stage in the disease and treatment pathway, and indicates which organisation(s) should be responsible for ensuring the patient has access to that information.

Table 4. The information pathway

Time	Nature of information	Responsibility
On referral to diagnostic clinic	Information on diagnostic clinic, tests it undertakes and who will be involved with the patient	Diagnostic clinic (see Chapter 3) by post
If sarcoma is suspected and the term is specifically used with the patient	Generic information on sarcoma	Diagnostic clinic
On diagnosis	Generic information on sarcoma. Specific information on the diagnosis (histological type, grade etc.) and the proposed treatment (if known)	Diagnostic clinic face-to-face or by telephone/post if requested by patient
Confirming referral to sarcoma treatment centre	Information on sarcoma treatment centre, names of consultants/nurses who will be involved in treatment and the named key worker for the patient Specific information on the diagnosis and the proposed treatment (if known and if not given by diagnostic clinic)	Sarcoma treatment centre (see Chapter 5) by post Local arrangements can apply
On any treatment recommendation	Generic information on that treatment (surgery, radiotherapy, chemotherapy) and any tests or imaging procedures that may accompany it. (Local or nationally published booklets may be appropriate)	Sarcoma treatment centre by post or face-to-face as appropriate
On referral to another sarcoma treatment centre	Reasons for the referral Information on the new sarcoma treatment centre. Identification of key worker	Referring sarcoma treatment centre face-to-face or by post New sarcoma treatment centre by post

Table 4. The information pathway *continued*

Time	Nature of information	Responsibility
After surgery or other treatment	<p>Specific information on individual follow-up procedure, self-monitoring information, healthcare support and sarcoma-specific support</p> <p>Confirmation of the named key worker for that patient together with contact details</p> <p>Specific information on support for prosthetic limbs or endoprosthetic implants</p> <p>Details about relevant rehabilitation services including provision of mobility aids, home adaptations and referral to local rehabilitation services</p> <p>Details of generic local and national support groups and other support resources</p>	<p>Sarcoma treatment centre by post or face-to-face as appropriate</p> <p>Sarcoma treatment centre or patient support centre, face-to-face or by post</p>
If targeted therapy is proposed (e.g. imatinib for GIST)	Generic information on the therapy and the applicable condition. Specific information relevant to the patient's own condition	Sarcoma treatment centre face-to-face, with copies by post to GP
In the event of advanced disease (whether at diagnosis or later)	Specific information on the nature of the advanced condition. Generic information will also be appropriate when metastatic disease is diagnosed	Sarcoma treatment centre face-to-face
When a clinical trial is proposed	Generic information on clinical trials. Specific information on the proposed trial	Sarcoma treatment centre face-to-face. Further information may come from trials unit by post
When no treatment other than palliative is available	Generic information on palliative care and pain control	Sarcoma treatment centre/palliative care centre face-to-face and GP

Generic information may include publications from national cancer charities and other voluntary sector providers, and this should be provided by the diagnostic clinic/sarcoma treatment centre.

When an existing clinical trial is not being conducted at the patient's own treatment centre, participation in that trial should be offered to the patient at another treatment centre.

Details of clinical trials for sarcoma should be available at every sarcoma treatment centre (see the recommendations on research in Chapter 10 'Improving knowledge').

Support

Patients and their carers should be offered appropriate support as follows:

- psychological support
- spiritual support
- social support through contact with others facing similar situations – self-help groups
- practical healthcare support relating to treatment
- benefits advice.

The development of sarcoma-specific self-help groups should be encouraged.

Patients should be supported in providing feedback to the sarcoma multidisciplinary team (MDT) to aid understanding of their service and patient needs, and to institute any changes.

General

Sarcoma treatment centres should collaborate so that duplication of resources to develop patient information leaflets/packs, Internet sites, information for GPs, etc. is minimised.

In the event of a delay or alteration in diagnosis that affects the management of a patient's condition, a 'significant event analysis' should be undertaken and the lessons learnt from this should be fed back to both relevant clinicians and MDTs. The patient should be informed by a senior doctor with appropriate skills (as defined in Chapters 3 and 4 of the NICE guidance on 'Improving supportive and palliative care for adults with cancer'⁷).

⁷ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

B. Anticipated benefits

Provision of clear, well-communicated and timely information will improve the patient's understanding of their condition and treatment, decrease their anxiety, and enhance their satisfaction with their care. This should increase overall compliance with care and may improve clinical outcomes.

Patients may benefit by both helping and being helped by others with a similar condition.

There will be a clear identification of what information should be provided, by whom and when.

C. Evidence

Communication

Evidence on techniques to improve communication between patients and healthcare professionals is reviewed in the NICE guidance on 'Improving supportive and palliative care for adults with cancer'.⁸ The benefit of communication skills training for healthcare professionals is supported by a systematic review. Evidence from three randomised controlled trials and one observational study supports the usefulness of a taped or written copy of consultations for patients.

Information

The development and distribution of information for patients and carers is considered in the NICE guidance on 'Improving supportive and palliative care for adults with cancer'.⁸ A systematic review confirms that patients with cancer obtain benefit from accurate information tailored to their diagnosis, stage and level of understanding.

Consistent, but limited, evidence from observational studies suggests that much Internet information on sarcoma is of poor quality, and contains inaccuracies.

⁸ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

In a survey by Sarcoma UK in 2004, 69% of the 45 respondents said they had looked for information about sarcoma on the Internet. Few patients said they had been offered general information about sarcoma during their treatment, although patients rated highly the information about their own situation given to them by doctors.

In another recent UK survey of a group of teenagers and young adults with cancer, 7% of whom had soft tissue sarcoma and 19% 'bone cancer', 46% replied that the cancer information they received was not appropriate for their age group.

Psychological and psychosocial support

Evidence from three systematic reviews considered in the NICE guidance on 'Improving supportive and palliative care for adults with cancer'⁹ suggests that psychosocial interventions are useful for the reduction of anxiety in people with cancer. The 2004 Sarcoma UK survey found that while patients with sarcoma were not routinely offered formal psychosocial support, those who attended counselling found it useful.

In two small observational studies, peer support programmes were viewed positively by participants with sarcoma. Many patients reported decreased anxiety and depression following the interventions. Only 15% of the respondents to the Sarcoma UK survey had attended a sarcoma-specific patient support group, although nearly half had some contact with other patients with sarcoma, usually during hospital clinics. While the majority of patients rated contact with their peers as positive, 9% found interaction with other patients difficult.

Patient travel

Evidence from one systematic review and seven observational studies suggests that while many patients find travelling to cancer treatment centres inconvenient, most are prepared to travel.

Evidence relating distance of travel to patient outcomes is inconclusive, because of the scarcity and heterogeneity of studies on this topic.

There is evidence from two American observational studies that when patients are presented with hypothetical treatment scenarios, some are prepared to accept an increased risk of morbidity or mortality in order to receive treatment in a local hospital.

⁹ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

D. Measurement

Structure

- provision of appropriate and adequate verbal and written information about the patient's diagnosis, proposed treatment options and sources of practical help
- provision of training courses in communication skills for the relevant healthcare professionals
- provision of self-help groups and other forms of support
- provision of a named key worker

Process

- evidence that patients receive appropriate and timely information
- evidence about the proportion of staff involved in patient care who have received formal communication training.
- surveys of patient experiences of each component of patient-centred care

Outcome

- improvement in patient experience as a result of action taken

E. Resource implications

Diagnosis

The resource implications of allocating a key worker to each patient are covered in the costs for staffing a sarcoma treatment centre. These costs are presented in Chapter 5 and are based on the key worker role being undertaken by a sarcoma clinical nurse specialist (CNS). In practice, however, any member of the sarcoma MDT may take on this role.

The total cost of offering every patient diagnosed with sarcoma in England and Wales a written and audio record of their consultation at diagnosis is estimated to be £1604 per year.

In Wales the resource implications for all patients to receive an audio and a written permanent record of their diagnosis would be £1150. In England, for all patients with sarcoma to receive an audio record of the consultation at diagnosis the cost would be £454; this assumes that these patients would already have funds allocated for written records.

Information

The total costs in England and Wales of producing information leaflets for patients with sarcoma are estimated to be £26,420 for the first year and £17,300 for subsequent years. This is for up to eight generic leaflets on different types of sarcoma and diagnostic clinic/sarcoma treatment centre-specific leaflets. This calculation assumes that the leaflets are designed and produced collaboratively to avoid unnecessary duplication.

Support

The resource implications of the recommendations on support for patients with sarcoma and their carers will require investigation by commissioners at treatment centre level. It may be helpful for commissioners to refer to the economic review for the NICE guidance on 'Improving supportive and palliative care for adults with cancer'.¹⁰

General

It is likely that there will be resource implications in the event of a significant event analysis being conducted as a result of a delay or alteration in diagnosis that affects patient management. This will require consideration on a case-by-case basis within the context of a multidisciplinary team.

¹⁰ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

Improving diagnosis of bone and extremity soft tissue sarcoma

3

The overriding principle is that any patient with a suspected or possible sarcoma needs to follow a clear and rapid pathway to diagnosis, and those with a confirmed sarcoma need to be referred promptly to a sarcoma treatment centre (see Chapter 5) for further management.

Public awareness of sarcomas is low and many studies have shown that some patients wait a considerable time after the onset of symptoms before seeking medical advice.

Because of their rarity, bone and soft tissue sarcomas are frequently difficult to diagnose and are characterised by late presentation and delays in diagnosis.

Extremity, trunk, and head and neck soft tissue sarcomas

For soft tissue sarcomas the principal problem in diagnosis is the large number of benign soft tissue tumours that cannot reliably be distinguished from malignant tumours (sarcomas) using clinical judgement.

The clinical guidelines (NICE ‘Referral guidelines for suspected cancer’¹¹) have defined the urgent referral criteria for soft tissue sarcomas and these may help to improve diagnostic accuracy. But, despite this, only one in ten referrals of ‘suspicious lumps’ will be a sarcoma. Therefore there is a large diagnostic workload that has to be addressed. Current practice and service provision generally fail to address this need, and this contributes to delay and adverse outcomes for patients who do have a malignant tumour. Currently diagnostic services for patients with these ‘suspicious’ soft tissue lumps are patchy, with few well-defined diagnostic clinics outside the major treatment centres.

¹¹ National Institute for Health and Clinical Excellence (2005) Referral guidelines for suspected cancer. *NICE clinical guideline* no. 27. Available from: www.nice.org.uk/CG027

Features suggestive of malignancy in a lump include:

- lump >5cm
- lump increasing in size
- lump deep to the fascia
- pain.

NICE 'Referral guidelines for suspected cancer' (www.nice.org.uk/CG027)

Patients with soft tissue lumps that do not meet the urgent referral criteria will not require referral to a diagnostic clinic.

Bone sarcomas

Patients with bone sarcomas often present to primary care with no palpable abnormality, and their symptoms are often very non-specific. The symptoms of malignant bone tumours cannot be reliably distinguished from a number of benign and self-limiting conditions. The diagnosis of a malignant bone tumour relies upon timely referral of the patient for an X-ray and recognition of the abnormality on the X-ray.

Because of the rarity of bone tumours, failure to recognise an abnormality on an X-ray or failure to identify it as being a tumour frequently contributes to the diagnostic delay for patients with bone sarcomas. Access to expert opinion to interpret abnormal X-rays is likely to be highly effective in triaging patients with abnormal X-rays and deciding what further investigations are required and where these should be carried out.

A. Recommendations

Referral guidelines

Commissioners should consider methods of increasing public awareness of the signs and symptoms of worrying lumps and the consequent need to attend a GP.

Commissioners should ensure that GPs are aware of and comply with the urgent referral criteria in the NICE 'Referral guidelines for suspected cancer'.¹²

¹² National Institute for Health and Clinical Excellence (2005) Referral guidelines for suspected cancer. *NICE clinical guideline* no. 27. Available from: www.nice.org.uk/CG027

Networks should ensure that GPs and hospital doctors are aware of the diagnostic pathways for patients with signs and symptoms suggestive of bone or soft tissue sarcoma.

Referral pathways: patients with extremity, trunk, and head and neck soft tissue sarcomas

To improve the early diagnosis of soft tissue sarcomas, a clearly defined network of diagnostic clinics linked to sarcoma treatment centres (see Chapter 5) should be established. Two models are recommended to achieve this:

either

1. patients with a suspected diagnosis of soft tissue sarcoma (as defined by the urgent referral criteria) would be seen within 2 weeks at a diagnostic clinic that is part of a sarcoma treatment centre

or

2. patients with a suspected diagnosis of soft tissue sarcoma (as defined by the urgent referral criteria) would be seen within 2 weeks at a diagnostic clinic specifically designated by their local cancer network. This would be a purely diagnostic, rather than a treatment clinic, and would be clearly affiliated to one sarcoma MDT (see Chapter 5).

Anyone with a possible sarcoma should be referred to a diagnostic clinic for biopsy. Biopsy should not be done outside these clinics.

Each cancer network should designate a diagnostic clinic for their patients who meet the urgent referral criteria. This would either be part of a sarcoma treatment centre or established locally, as described above.

The diagnostic clinics (in either model) should undertake triple assessment including clinical assessment, imaging and biopsy of all patients. There would be no requirement for a surgeon or oncologist to be part of such a team, but the members of the diagnostic team should be trained by and work in close collaboration with members of the affiliated sarcoma MDT. Patients identified as having a soft tissue sarcoma should be rapidly referred on to a sarcoma MDT for definitive treatment, as would any cases with equivocal images or biopsy.

A diagnostic clinic separate from a sarcoma treatment centre should have its staff trained and its work audited by the sarcoma MDT from the sarcoma treatment centre to which it is affiliated.

Appropriate imaging facilities should be available to comply with national access standards (as defined in the 'NHS Cancer Plan'¹³ and the 'Wales National Cancer Standards'¹⁴).

Some patients with a soft tissue sarcoma will be diagnosed following excision of a lump thought to be benign but which turns out to be malignant. These patients should be referred directly to the sarcoma MDT designated by that cancer network.

Patients whose lump turns out to be benign should be referred locally for appropriate management.

Commissioners and networks should work together to ensure that there are clear referral pathways from both primary and secondary care through to a designated diagnostic clinic and for patients with proven sarcomas on to the affiliated sarcoma treatment centre.

An audit of all elements of the referral pathway should be carried out.

Referral pathways: bone sarcomas

All patients with a probable bone sarcoma (usually following X-ray examination) should be referred directly to a bone tumour treatment centre (see Chapter 6) for diagnosis and management.

Appropriate imaging facilities should be available to comply with national access standards (as defined in the 'NHS Cancer Plan'¹³ and the 'Wales National Cancer Standards'¹⁴).

The biopsy of patients with a possible bone sarcoma should only be carried out at a bone tumour treatment centre.

Patients with X-ray abnormalities that are most likely to be due to a secondary malignancy or a benign process should be referred to the local orthopaedic service for further investigation. Networks should consider formalising service provision for this latter group.

An audit of all elements of the referral pathway should be carried out. Some patients with a bone sarcoma will be diagnosed following surgery. These patients should be referred directly to the sarcoma MDT designated by that cancer network.

¹³ Department of Health (2002) *The NHS Cancer Plan*. Available from: www.dh.gov.uk (accessed 3 January 2006)

¹⁴ Welsh Assembly Government (2005) *Wales National Cancer Standards*. Available from: www.wales.gov.uk/subihealth/content/cancer/national-standards-e.htm

Radiology review

If a plain X-ray shows abnormalities that could be a bone sarcoma, there should be clear arrangements for review of these images by specialist sarcoma radiologists at a sarcoma MDT. This service should be recognised and funded appropriately.

Histopathology review

All patients with a possible diagnosis of bone or soft tissue sarcoma should have the diagnosis confirmed by a specialist sarcoma pathologist (see Chapter 4).

B. Anticipated benefits

There will be a clear referral pathway from primary and secondary care to the appropriate sarcoma MDT and this will reduce delays in diagnosis.

Clearly identified services for diagnosis and treatment of sarcoma will result in an increased proportion of patients diagnosed and treated by recognised specialists; this should lead to improvements in patient care and outcomes.

Review by specialist pathologists of all tissue samples thought to be sarcoma will improve diagnostic accuracy.

Radiological review of abnormal imaging and guidance on further investigation will avoid unnecessary imaging.

C. Evidence

Patient-related delay in diagnosis

Some patients with sarcoma wait a considerable time after the onset of symptoms before seeking medical advice. Several observational studies have reported such patient-related delay in sarcoma.

In a Belgian study, 47% of patients with soft tissue sarcoma showed delay of more than 1 month in seeking medical advice. The median delay in this subgroup was 4 months. In a Dutch study, 36% of patients with retroperitoneal soft tissue sarcoma waited for more than 6 months following the onset of symptoms before seeing a doctor.

The shortest patient delays were reported for patients with osteosarcoma; in five studies estimates of mean or median delay ranged from 1 to 1.6 months. Patient delays were longer for those with Ewing's sarcoma; estimates of mean or median delay from six studies ranged from 1.5 to 4 months.

Referral delay

Diagnostic uncertainty at the point of consultation to primary or secondary care can result in a delay in referral to the appropriate treatment centre. Several observational studies reporting referral delay were identified.

In a study of referral to a UK specialist soft tissue sarcoma unit, delay of more than 3 months was seen in 20% of cases. Median delay in this subgroup was 14 months. The most frequent reason for delay was lack of clinical suspicion at the initial consultation. A second UK study reported referral delay of patients with malignant bone or soft tissue tumours to a specialist treatment centre. On average, referral to the treatment centre from the patient's GP or local hospital took 7.5 months.

An observational study, using data from the Northern and Yorkshire Cancer Registry, examined referral patterns for 362 patients with non-gynaecological sarcoma in the years 1999–2000. Only 60% of these patients were eventually referred to a specialist sarcoma treatment centre, many experiencing considerable delay in the process.

In a recent UK survey of a group of teenagers and young adults with cancer, 42% of those with soft tissue sarcoma said they visited their GP more than five times before they were referred to hospital. The mean number of physician visits before referral to a specialist unit for bone or soft tissue sarcoma was 4.85 in an American study.

Several observational studies reported the interval from the first consultation with a doctor to the eventual diagnosis of sarcoma (the doctor-related diagnostic delay). Estimates of doctor-related diagnostic delay were shortest for osteosarcoma, ranging from 1.2 to 2.25 months in five papers. Longer doctor-related diagnostic delays were reported for patients with Ewing's sarcoma of bone (1.25 to 7.75 months; five studies). One American study found that 44% of patients with primary pelvic bone sarcomas experienced a doctor-related diagnostic delay greater than a month. Median delay was 7 months in this group.

A large American study reported that 50% of patients with soft tissue sarcoma experienced a doctor-related diagnostic delay of 2 months or more and in 21% of cases delay was more than 6 months. Similarly in a Belgian study, doctor-related delay of more than 1 month was seen in 27% of patients with soft tissue sarcoma. The median delay in this subgroup of patients was 6 months.

Diagnostic delay and outcome

Evidence relating diagnostic delay to patient outcomes in sarcoma was limited in quantity, and observational in nature. The studies tended to include a mixture of cases, making it difficult to estimate the prognostic significance of delay.

Several studies expressed the opinion that the increase in tumour size during a delay in diagnosis has a detrimental effect on treatment options and outcomes in patients with sarcoma.

In a UK study of patients with soft tissue sarcoma, which was partially adjusted for case mix, preoperative duration of symptoms for more than a year was associated with better survival. This suggests diagnostic delays may be a feature of lower grade tumours. This notion is supported by four other studies of patients with bone and soft tissue tumours, which found patient-related and referral delays tended to be longest for patients with benign tumours and shortest for those presenting with metastatic disease.

One study of a Scandinavian soft tissue sarcoma treatment centre attributed a historical improvement in local control and survival to better referral practices. This meant more patients presented with small subcutaneous lesions and better prognosis.

Two studies reported adverse outcomes in patients who had been misdiagnosed and managed inappropriately. In a series of patients with musculoskeletal tumours that had been misdiagnosed, 60% of cases required a more radical surgical procedure than would originally have been necessary, because of diagnostic delay or contamination of the tumour margins. A Dutch population-based study of retroperitoneal soft tissue sarcoma reported that complete resection of the tumour was less likely in patients with a preoperative misdiagnosis than in those in whom the diagnosis of sarcoma was considered. This was partly because unnecessary surgery for an inoperable tumour was more likely in those with preoperative misdiagnosis.

Pre-referral imaging

Evidence from two observational studies suggests that ordering of a radiograph by the primary care physician is associated with reduced diagnostic delay in suspected primary bone tumours.

Three observational studies of pre-referral magnetic resonance imaging (MRI) or computed tomography (CT) imaging in primary bone and soft tissue tumours were found. MRI or CT imaging was often technically inadequate and had to be repeated, contributing to a delay in diagnosis. One study noted a tendency in referring centres to perform too many MRI sequences.

Radiological diagnostic service

A UK observational study found that 19% of bone tumours referred to a treatment service had been missed by both the clinician and the radiologist on the initial radiograph, though the tumour was evident on retrospective review of the image. In the group of patients whose initial radiographs were erroneously reported as normal, diagnostic delay meant that 58% required amputation or were inoperable compared to 15% of those whose initial radiographs were interpreted correctly.

There is consensus among radiologists working in the specialist areas that specialist review of the imaging in people with suspected sarcoma reduces clinical error rates and delay in diagnosis.

An audit of specialist oncological radiology review in a UK cancer centre (6% of the patients had sarcomas) noted that while cross-sectional imaging was usually technically adequate there was often a difference in interpretation between specialist and general radiologists.

Biopsy

There is evidence from seven observational studies that a well-performed preoperative biopsy is more likely if a patient is referred to a specialist with an interest in sarcoma management. Four of these studies, one from the UK, one from the USA and two from Australia, also reported patient outcomes. Adverse outcomes attributed to poorly performed or planned biopsies included unnecessarily extensive surgical resection (including amputation) and local recurrence. Such outcomes were less likely when biopsy was performed by a specialist with an interest in sarcoma management.

Pathology

Evidence about histopathology review by an expert pathologist, obtained from observational studies, suggests that the diagnosis of sarcoma is often altered on expert review (see Chapter 4).

D. Measurement

A key factor in improving outcomes for patients with sarcoma is prompt referral to an efficient diagnostic pathway followed by management by a sarcoma MDT. The patient pathway will be routinely audited and reviewed under the national cancer peer review system and waiting times targets (as defined in the 'NHS Cancer Plan'¹⁵ and the 'Wales National Cancer Standards'¹⁶).

Structure

- provision of clear protocols and information on referral processes
- provision of adequate staff and resources for effective functioning of all specialist sarcoma MDTs in order to ensure compliance with waiting time requirements for diagnosis (as defined in the 'NHS Cancer Plan'¹⁶ and the 'Wales National Cancer Standards'¹⁵).
- provision of adequate access to appropriate imaging equipment and specialist radiologists
- sessional time and appropriate laboratory facilities for specialist histopathologists to attend specialist MDT meetings and provide a diagnostic service

Process

- compliance with NHS targets for diagnosis and treatment relevant to sarcoma services
- the percentage of patients referred to the sarcoma MDT before the first definitive treatment in order to monitor the frequency of non-specialist treatment

¹⁵ Department of Health (2002) *The NHS Cancer Plan*. Available from: www.dh.gov.uk (accessed 3 January 2006)

¹⁶ Welsh Assembly Government (2005) *Wales National Cancer Standards*. Available from: www.wales.gov.uk/subihealth/content/cancer/national-standards-e.htm

- the percentage of patients with sarcoma undergoing preoperative scan (MRI or CT) and staging investigations before first definitive treatment in order to monitor appropriate diagnostic management
- the percentage of patients with sarcoma undergoing preoperative diagnostic biopsy (excluding retroperitoneal sarcomas) before first definitive treatment
- survey of patient satisfaction

Outcome

- action taken as a result of patient satisfaction survey
- effect of diagnostic accuracy on patient outcomes

E. Resource implications

Referral guidelines

The resource implications of increasing public awareness of worrying lumps are beyond the scope of this guidance and have therefore not been costed.

There may be resource implications associated with the need to ensure GPs and hospital doctors are aware of the diagnostic pathways for patients with features suggestive of bone or soft tissue sarcoma. This cost has not been included in the economic review; the centre-specific leaflets produced to inform patients of their diagnostic and treatment pathway are likely to be of assistance to healthcare professionals.

Referral pathways: patients with extremity, trunk, and head and neck soft tissue sarcomas

The annual opportunity cost for each clinic operating one diagnostic session per week for 45 weeks of the year, inclusive of four training sessions, is estimated to be between £88,833 and £101,622. The variation depends upon whether the clinic is led by a doctor or by a sarcoma CNS, and on the level of activity. The costs presented here do not take into account the cost of any new equipment, such as ultrasound, that may be needed to provide this diagnostic service. These values represent the estimated maximum cost of staffing a new diagnostic clinic.

There are around 2000 patients newly diagnosed with soft tissue sarcomas each year in England and Wales, and it is estimated that around ten times this number will have been referred with suspicious lumps for diagnosis. If all these patients, 20,000, are referred to a diagnostic clinic for triple assessment, the total cost would be at least £4,260,000 per year (interquartile range £3,000,000 to £4,840,000).

Triple assessment for sarcoma is likely to be more expensive than for breast cancer. Approximately 30% of the 20,000 patients referred for diagnostic tests will require an MRI scan in addition to an ultrasound, to confirm their diagnosis. The average unit cost for an MRI scan is £223.94 (interquartile range £184 to £465), therefore the additional cost for this patient group would be approximately £1,343,640 in England and Wales (interquartile range £1,164,000 to £2,790,000). It should be noted however that in many diagnostic clinics triple assessment of patients with suspected sarcoma is already being carried out.

The provision of appropriate imaging facilities will have capital, operational and staffing cost implications where access is limited. This will need to be considered by local commissioners.

Referral pathways: bone sarcomas

Services relating to the diagnosis of bone sarcoma are funded by the NSCAG and Health Commission Wales, and have therefore not been considered separately here.

Bone tumours are rare tumours, and the majority of patients with a primary malignant bone tumour will be treated in a centre that deals with a significant volume of patients with this condition. As a result there are only a small number of pathologists who see significant numbers of bone tumours. There are approximately 20 benign soft tissue lumps removed for every one soft tissue sarcoma in a typical district general hospital. Most general pathologists report some soft tissue tumours. Primary soft tissue sarcomas are rare, and there are many subclassifications that are important for tailoring treatment. Bone and soft tissue sarcomas have many mimics, both benign tumours and reactive conditions, making this a particularly complex field of pathology.

Errors in diagnosing bone and soft tissue sarcomas are not uncommon in those pathologists who are not specialists in this field. These errors can be minimised by review of all suspicious histology by an appropriately experienced pathologist.

Increasingly, the use of cytogenetics and molecular pathology is becoming an essential tool in confirming the diagnosis of certain sarcomas, and may well have both treatment and prognostic implications. These facilities require funding at one or more centres. Storage of tissue samples is important for future research in these rare tumours.

GIST are also rare tumours, and in the majority of cases the diagnosis is only established after resection of the primary tumour. These tumours are usually referred either to pathologists with an interest in soft tissue tumours, or to pathologists with an interest in gastrointestinal tumours. The diagnosis is based on tumour morphology, knowledge of tumour site and a panel of immunohistochemical markers. In cases of doubt, mutation analysis may be helpful.

There is a serious shortage of consultant pathologists in the UK with up to 20% of consultant posts unfilled. There are recognised specialists in bone and/or soft tissue tumours and there is a group of pathologists who do general pathology but have a special interest and expertise in bone and/or soft tissue tumours.

These two groups comprise just 26 pathologists (26 in England and 0 in Wales) and are referred to in this guidance as specialist sarcoma pathologists (SSP). Together they form and take part in External Quality Assurance (EQA) schemes in bone and soft tissue pathology and form an informal network for slide and peer review. The diagnosis of GIST tends to be initiated locally, with the pathology material being reviewed by a soft tissue specialist or a specialist gastrointestinal (GI) pathologist.

The Department of Health now requires NHS pathology laboratories to enrol in a laboratory accreditation scheme. There is almost exclusive use of Clinical Pathology Accreditation (UK) Limited (CPA) in NHS laboratory accreditation. As part of that accreditation pathologists must participate in relevant EQA schemes and CPA does not normally accredit single-handed practice. As part of accreditation there must be documented audit.

A. Recommendations

All primary malignant bone tumours should either be first reported or reviewed by an SSP-bone. An SSP-bone is a pathologist who regularly reports bone tumours and these form a significant component of their workload. He or she should successfully participate in the bone part of the bone and soft tissue pathology EQA scheme, and be part of a properly constituted sarcoma MDT.

All soft tissue sarcomas should either be first reported or reviewed by an SSP-soft tissue. An SSP-soft tissue is a pathologist who regularly reports soft tissue tumours and these form a significant component of their workload. He or she should participate in the soft tissue part of the bone and soft tissue pathology EQA scheme and be part of a properly constituted sarcoma MDT.

All GISTs should be reported or reviewed by an SSP with experience in GIST who successfully participates in the bone and soft tissue pathology EQA scheme, or a tertiary GI specialist who successfully participates in the GI pathology EQA scheme.

All patients with soft tissue tumours assessed in a diagnostic clinic (see Chapter 3) should have their pathology reported by:

either

- an SSP-soft tissue

or

- a pathologist nominated by the sarcoma MDT as part of the local diagnostic referral pathway who has formal links to an SSP.

All malignant soft tissue tumours should be reviewed by an SSP-soft tissue prior to management recommendations by the sarcoma multidisciplinary team (MDT).

Pathology reports should include all the information required by the Royal College of Pathologists' histopathology dataset for soft tissue sarcomas once it is available. They should use a defined tumour classification (for example, the World Health Organization (WHO) classification 2002) and grading (for example, the Trojani grading system).

The Royal College of Pathologists should be asked to expedite production of a histopathology dataset for bone and soft tissue sarcoma, and should be invited to give guidance on situations where molecular diagnosis is of value.

There should be at least conditional CPA approval for the laboratory in which the SSP and those with a specialist interest work. There should be formal documented audit of the work of the SSPs and the nominated pathologists.

The SSPs should have ready access to molecular pathology and/or cytogenetics facilities.

All sarcoma MDTs (see Chapter 5) must have at least one, or ideally two, SSPs. Where there is only one SSP, formal links with an SSP in another centre should be established for the purposes of consultation, audit and cross-cover.

The additional work of reviewing cases by SSPs should be recognised in their job plan.

Commissioners should fund:

- a formal system for second opinions and review of difficult cases
- molecular pathology and cytogenetics facilities.

All pathology laboratories in centres treating bone or soft tissue sarcomas should store tissue in appropriate facilities for research (subject to the provisions of the Human Tissue Act).

Commissioners should consider funding sarcoma pathology fellowships to address the current shortages of SSPs.

B. Anticipated benefits

The use of specialist pathologists will reduce the risk of errors in diagnosis in primary bone and soft tissue sarcomas.

Work undertaken in an accredited laboratory will help ensure high-quality pathology services.

The establishment of a formal network between SSPs and nominated pathologists will help ensure accuracy in diagnosis, and encourage training and education.

A national tissue resource will be created by a large tissue bank of sarcoma material for future research.

C. Evidence

There is consistent evidence that the histopathological diagnosis of sarcoma is often changed on review by an expert pathologist. A recently audited UK referral practice for bone and soft tissue tumours (second opinions) showed that 9% of cases sent as some form of malignant tumour were benign or reactive conditions, and 6% of tumours sent as benign conditions were malignant. There was a discrepancy of 18% when it came to categorising tumour type, with 40% of these likely to affect prognosis and/or treatment. Evidence from 16 other observational studies supports these findings. Four of these studies included bone sarcomas only, eight soft tissue sarcomas only and four studies both bone and soft tissue sarcomas.

Nine studies reported the rate at which a diagnosis of sarcoma is changed to non-sarcoma on expert review. Estimates ranged from 3% to 22%.

Ten studies reported that the subtype of sarcoma was changed on expert review in between 16% and 39% of cases.

Six studies examined how often the expert pathologist disagreed with the tumour grade recorded in the original histopathological report; estimates ranged from 24% to 40%.

One study reported a lower diagnostic error rate at musculoskeletal tumour treatment centres (13%) than at referring institutions (24%).

Central histopathological review as part of the European Osteosarcoma Intergroup clinical trial found 2% of the patients randomised to participate were in fact ineligible as a result of incorrect pathology.

Evidence from observational studies and a systematic review suggests that cytogenetic or molecular pathology testing can identify characteristic genetic aberrations in some subtypes of sarcoma. This is relevant in the diagnosis of sarcomas that are difficult to distinguish histologically but where patient management depends on the diagnosis (for example, alveolar versus embryonic rhabdomyosarcoma; synovial sarcoma versus other spindle cell sarcomas or c-kit mutational analysis in GIST).

Consensus statements by European and American expert sarcoma pathologists support the clinical usefulness of genetic tests in the diagnosis of certain types of sarcoma.

D. Measurement

Structure

- availability of SSPs for both bone and soft tissue and ready access to molecular pathology/cytogenetics
- availability of second opinions

Process

- time from GP referral to definitive diagnosis
- time between biopsy and receipt of pathology report
- attendance of SSP at MDT meetings
- participation in an appropriate EQA scheme
- audit of results of nominated pathologists' work

Outcome

- effect of diagnostic accuracy on patient outcomes
- patient satisfaction

E. Resource implications

The recommendations that all primary malignant bone tumours, soft tissue sarcomas and GISTs should be reported or reviewed by a specialist sarcoma pathologist will increase the referral rates and workload. In view of the current shortage of pathologists, any additional staffing requirement may not be immediate.

It is likely that there would need to be an additional clinical scientist, biomedical scientist and secretary employed at each of the four existing cytogenetic/molecular pathology laboratories. The additional annual employment cost per existing laboratory per year would be approximately £79,950.

Improving treatment: sarcoma multidisciplinary teams

Multidisciplinary teams (MDTs) have become the accepted way of delivering modern cancer care, and such teams are often complex with varying membership, depending on the location of care and the tumour type. MDTs need a minimum number of patients to maintain expertise and justify the resources required for their support.

The Guidance Development Group considered the evidence for the minimum number of patients a sarcoma MDT should serve. Justification for the numbers chosen is based on the minimum number of patients necessary to justify establishment of a sarcoma MDT and to maintain skills. In practice, a soft tissue sarcoma MDT is likely to serve a population of 2–3 million people and a bone sarcoma MDT one of 7–8 million.

The development and support for such a team need to be properly recognised within the management structure of a trust, with the identification of a clinical lead with appropriate responsibility, authority and access to resources.

The team needs to take responsibility for both the diagnostic pathway and treatment of all patients with sarcoma within their catchment area. This, in effect, creates a managed sarcoma network – clinicians, commissioners and cancer networks need to work together to identify the catchment population served and develop the appropriate diagnostic and treatment pathway to serve their population.

The appropriate care of patients with less common bone and soft tissue sarcomas is more complicated. More than one MDT will need to consider the management of the patient. These include head and neck, gastrointestinal and uterine teams. Particularly complex cases may need referral to a more experienced MDT for advice.

Because of the rarity of primary malignant bone tumours, NSCAG has taken on the commissioning of diagnostic and surgical services for this group of patients. An equivalent commissioning process does not exist for soft tissue sarcoma.

A. Recommendations

All patients with a confirmed diagnosis of bone sarcoma, or adults with a soft tissue sarcoma, should have their care supervised by or in conjunction with a sarcoma MDT.

The sarcoma MDT should be expected to manage at least 100 new patients with soft tissue sarcoma per year; if the MDT also manages bone sarcomas then it should manage at least 50 new patients with bone sarcoma plus 100 new patients with soft tissue sarcoma.

This guidance should be implemented by primary care trusts (PCTs)/local health boards (LHBs) working collaboratively through their specialist commissioning groups, in close consultation with cancer networks. A National Implementation Group should be considered for both England and Wales.

Each sarcoma MDT should be based either in a single hospital or in several geographically close and closely affiliated hospitals, which would constitute the sarcoma treatment centre.

There should be a nominated clinician (clinical lead) who takes responsibility for the service and this should be reflected in their job plan. The clinical lead should be a member of the core MDT.

Information about the specific expertise of different MDTs should be made widely available so that cases can be referred expeditiously (see Chapter 7). Such expertise – which is not likely to be found everywhere – includes:

- gynaecological sarcomas
- head and neck sarcomas
- retroperitoneal and pelvic sarcomas
- chest wall/intrathoracic sarcomas
- skin sarcomas
- central nervous system sarcomas
- gastrointestinal stromal tumours (GIST)
- adult-type soft tissue sarcomas arising in children
- the use of isolated limb perfusion.

Sarcoma MDT membership

Each sarcoma MDT should have a core membership as shown in Table 5.

Table 5. Core membership of a sarcoma multidisciplinary team

Staff requirements	Specification
Specialist sarcoma surgeon	A minimum of two per MDT. These surgeons should have a major clinical interest in sarcomas i.e. spend at least 5 programmed activities of direct clinical care involved in managing sarcomas
Specialist sarcoma radiologist	At least two with a special interest in musculoskeletal/oncological imaging
Specialist sarcoma pathologist	At least one and ideally two (see Chapter 4)
Medical oncologist and/or clinical oncologist	At least two with an interest in musculoskeletal oncology. There should be at least one clinical oncologist. The oncologist/s should each spend a minimum of three programmed activities of direct clinical care involved in the management of sarcomas
Sarcoma clinical nurse specialist/key worker*	Sufficient to allocate a clinical nurse specialist/key worker for each patient (but a minimum of two) – see Chapter 8
Support staff	MDT coordinator and secretarial support
Palliative care specialist	A member of the specialist palliative care team

* Key worker may come from any of the disciplines involved in the multidisciplinary team (MDT).

Each MDT should in addition have an extended team with membership as shown in Table 6, some of whom (for example key workers) may work as part of the core team.

Table 6. Membership of an extended sarcoma multidisciplinary team

Staff requirements	Specification
Specialist sarcoma physiotherapist	With expertise in sarcomas
Specialised allied health professionals (AHP)	Consisting of other relevant AHPs, such as therapy radiographers, occupational therapists, prosthetists, orthotists, dietitians and social workers, plus access to counsellors and/or psychologists
Paediatric oncologist	Specifically for MDTs that treat children and young people with bone and/or soft tissue sarcoma
Specialist nurse(s)	Including palliative care nurses and appropriately trained ward staff
Affiliated medical or clinical oncologist from linked cancer centre	Nominated by the cancer network clinical director and approved by the MDT lead clinician
Affiliated diagnostic service clinicians	Nominated by the cancer network clinical director and approved by the MDT lead clinician
Other professionals including orthopaedic, thoracic, plastic, head and neck, gynaecological, GI and vascular surgeons	Nominated by the cancer network clinical director and approved by the MDT lead clinician

Members of the extended team should be nominated and will bring particular expertise to the sarcoma MDT. They should attend MDT meetings as and when appropriate.

Role of the sarcoma MDT

The MDT should:

- have weekly meetings at which all core members of the team are present and their attendance is documented
- ensure that a treatment plan is agreed and documented by the MDT for all of the following:
 - newly diagnosed patients
 - patients following tumour resection
 - patients with first metastases and/or first local recurrence

- ensure that the written care/treatment plan draws together the provision of all components of care
- ensure that a key worker has been allocated to each patient
- cooperate in service development at a national and local level for patients with sarcomas
- ensure national standards for diagnosis and treatment (as defined in the 'NHS Cancer Plan'¹⁷ and the 'Wales National Cancer Standards'¹⁸) are achieved
- have operational policies for the diagnosis and treatment of patients
- have documented arrangements for linking with other MDTs to ensure coordinated management of patients with sarcomas at specific anatomical sites for which specialist input is required (for example, head and neck, uterine, retroperitoneal sarcoma and GIST; see Chapter 7)
- comply with the information requirements of the National Cancer Dataset
- participate in any future national audit programmes for sarcoma outcomes
- participate in national and international trials
- ensure audit and education of its referring hospitals and networks
- ensure GPs are given prompt and full information about significant changes in their patients' illness or treatment
- encourage education of medical students, GPs and trainee surgeons about the diagnosis and management of sarcomas.

¹⁷ Department of Health (2002) *The NHS Cancer Plan*. Available from: www.dh.gov.uk (accessed 3 January 2006)

¹⁸ Welsh Assembly Government (2005) *Wales National Cancer Standards*. Available from: www.wales.gov.uk/subihealth/content/cancer/national-standards-e.htm

B. Anticipated benefits

All patients will be seen and assessed rapidly at a centre with appropriate expertise.

All aspects of the patients' management will be consistent and well coordinated, with appropriate use of specialist skills.

Coordinated specialist rehabilitation and supportive care will be provided.

National programmes will be supported and encouraged.

The sarcoma treatment centre will be the focus of continual improvements to the pathways of care for patients served by the centre.

More patients will be entered into clinical trials.

C. Evidence

It is difficult to separate the effects of MDTs, specialist centres and hospital case volume on patient outcomes. Multidisciplinary sarcoma teams tend to be located in specialist centres which in turn treat the greatest numbers of patients. The rarity of sarcomas means a lack of research originating in primary or secondary care, so the evidence is restricted to case series from tertiary and quaternary centres or studies using cancer registries. Pretreatment differences between patients cared for by specialist and non-specialist centres could confound comparisons between the two settings, but few studies adjusted for case mix.

MDTs for bone sarcoma

Evidence relating management by MDT for people with bone sarcomas was limited to a single UK cohort study of patterns of care and survival in patients younger than 40 years with bone sarcoma, which partially adjusted for case mix. Patients managed by a specialist MDT at the supraregional bone tumour services or United Kingdom Children's Cancer Study Group (UKCCSG) paediatric oncology centres in England and Wales had better overall survival when compared to those treated at other hospitals. The study was not designed to address the issue of MDT management and it is unknown whether any of the other hospitals had MDTs treating people with bone sarcomas.

MDTs for soft tissue sarcoma

There was consistent evidence from observational studies in favour of specialist MDTs for the management of patients with soft tissue sarcoma. The studies used cancer registries and hospital records to compare the outcomes of patients treated in different settings. Four studies, two from Scandinavia and one each from Canada and the UK, included only patients with soft tissue sarcoma of the limb, limb girdle or trunk. A French audit contained a majority of patients with extremity or truncal soft tissue sarcoma, but also some patients with soft tissue sarcoma at other anatomical sites. The UK study was the only one to adjust for differences in case mix in its analyses.

There was evidence of an overall survival advantage for those patients with soft tissue sarcoma treated by a sarcoma MDT in the three studies that reported this outcome. The four studies that considered disease-free survival found an advantage for those patients who were treated by a sarcoma MDT.

None of the three comparisons of surgical resection margins were case-mix adjusted. Two studies reported that wide or compartmental surgical resections were more likely for patients treated by a sarcoma MDT. The UK study did not observe a difference between the rate of wide or compartmental resections achieved by the sarcoma MDT and by district general hospitals in the same region, although 45% of the patients treated by the MDT had large, high-grade, deep sarcomas, compared to 21% of those treated at district general hospitals. Differences in the determination of surgical margins between centres may confound comparisons. An American observational study of patients with soft tissue sarcoma noted that in non-specialist treatment centres 59% of surgical resections reported as 'wide' were found to contain residual disease on specialist pathological review.

Other differences between patterns of care provided by specialist sarcoma MDTs and those provided by other treatment centres included better conformity to clinical practice guidelines by MDTs and greater use of preoperative imaging and biopsy.

Hospital case volume and patient outcome

There is consistent evidence, reviewed for example in NICE guidance on 'Improving outcomes in colorectal cancers',¹⁹ that in complex or high-risk surgery for cancer, case volume appears positively associated with improved patient outcomes.

¹⁹ National Institute for Clinical Excellence (2004) Improving outcomes in colorectal cancers. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=204541

Evidence about hospital case volume and outcome in patients with sarcoma was limited to two observational studies and a cohort study. Because of the rarity of sarcoma, definitions of ‘high case volume’ tended to be generous, ranging from one patient per year to ten or more patients per year.

The UK bone tumour cohort study examined the effect of hospital case volume on the survival of patients with osteosarcoma or Ewing’s sarcoma. Hospitals were categorised according to the average number of new patients treated per year: 0–1, 2–4, 5–9 and more than 9 patients. Partial adjustment for case mix was made in the analysis. A beneficial effect of hospital case volume on survival was observed for patients with Ewing’s sarcoma but not for those with osteosarcoma.

A Dutch observational study compared the outcomes of patients with retroperitoneal soft tissue sarcoma in hospitals treating an average of fewer than one patient per year with those in hospitals treating more than one patient per year. Adjustment was made for case mix. While complete resection of the tumour was more likely in the higher-volume hospitals, no effect on survival was observed. Another Dutch study noted that adherence to guidelines for the diagnosis of soft tissue sarcoma was more likely in hospitals treating more than two patients per year.

A Canadian observational study of patients with extremity soft tissue sarcoma compared patient outcomes in three categories of hospital case volume: fewer than 2, 2–5 and more than 5 cases per year on average. The case volume of the hospital providing definitive treatment was not statistically associated with risk of amputation or survival. No adjustment for case mix was made in this study.

D. Measurement

Structure

- evidence that sarcoma MDTs have been established with the formal agreement of the cancer networks
- sarcoma MDTs are staffed appropriately
- evidence for clear arrangements for diagnosis, whether at a diagnostic clinic which is part of the sarcoma treatment centre or at a specifically designated diagnostic clinic in a local cancer network

Process

- evidence that the sarcoma MDT manage the minimum number of patients as defined in this guidance
- every patient is managed by the appropriate MDT
- participation by individual specialists in MDT meetings
- audit and review by surgeons and histopathologists
- all members of the sarcoma MDT participate in continuing professional development (CPD) or a personal development plan (PDP)
- operational policies for referral of patients to the centres
- demonstration of links to appropriate specialist surgical expertise

Outcome

- improvements of clinical outcomes
- increase in clinical trial participation
- patient satisfaction

E. Resource implications

The resource implications of this section are based on minimum staffing levels for sarcoma treatment centres to provide a safe and sustainable service. The annual employment costs of the medical, nursing and other staff caring for 100 new patients per year is estimated to be between £482,399 and £819,039. These costs represent opportunity costs because the staff involved in the treatment centres are likely to already be contracted to the NHS.

There is likely to be an additional requirement for some healthcare professionals, in particular sarcoma CNS and specialist sarcoma physiotherapists. This needs to be considered by commissioners.

The resource implications of the National Implementation Group, including staff employment costs and expenses for board members, have been estimated to be between between £98,443 and £100,843 per annum.

Improving treatment: bone sarcomas

The key factors that influence the provision of services for patients with primary bone sarcoma are:

- the low incidence of cases (400 cases per year in England and Wales)
- the young age of many of the patients
- the complexity of treatment
- requirements for rehabilitation.

In view of the young age of many of these patients, the recommendations in the NICE guidance on 'Improving outcomes in children and young people with cancer',²⁰ should be observed.

NSCAG has long recognised the complexity of the surgical treatment of bone tumours, and the majority of surgical procedures are carried out in NSCAG-recognised bone tumour treatment centres. In operable cases, limb salvage can be achieved in 85% of patients; the remainder will need amputation.

Chemotherapy regimens used for bone sarcoma are among the most complex in adult oncology practice. About 75% of patients with bone sarcoma will need chemotherapy, of whom 60% will be under the age of 20 years. This means that 300 patients with bone sarcomas need chemotherapy, of whom 180 are aged under 20 years.

Chemotherapy can have life-threatening toxicity (for example, neutropenic sepsis) and is associated with an increased risk of second malignancy. Given that patients are often young when treated and then become long-term survivors, the issue of fertility preservation and other late effects is particularly important.

²⁰ National Institute for Health and Clinical Excellence (2005) Improving outcomes in children and young people with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

Radiotherapy is a key part of treatment for many patients with Ewing's sarcoma (70%), approximately 60 patients per year in England and Wales. It is an important and valuable part of palliative therapy for other patients with bone sarcoma. Radiotherapy is typically delivered by fractionation of the total dose over 4 to 6 weeks with daily attendances for treatment.

A. Recommendations

Surgery

All patients with bone sarcoma should undergo definitive surgical resection at a bone tumour treatment centre with a properly constituted MDT.

A bone sarcoma MDT should see a minimum of 100 new cases of bone sarcoma per year (or 50 cases of bone sarcoma if the MDT also manages 100 cases of soft tissue sarcoma).

Chemotherapy and radiotherapy

There should be a formal relationship between the bone sarcoma MDT and the provider of non-surgical oncology services that is characterised by common protocols, good communication, and well-defined referral pathways. These relationships should be defined in writing and approved by the cancer network director and the bone sarcoma MDT lead clinician. Audits of compliance with these protocols will need to be demonstrated.

The provider of chemotherapy services should:

- a) provide the facilities for intensive inpatient chemotherapy as described in the 'Manual for cancer services' 2004²¹
 - b) be **either**
 - a principal treatment centre for children or young people (likely to be a UKCCSG Centre or a teenage cancer unit)
- or**
- an adult cancer centre with a formal relationship with a bone sarcoma MDT

²¹ Department of Health (2004) *Manual for Cancer Services*. Available from: www.dh.gov.uk (accessed 3 January 2006)

- c) have a clinical/medical oncologist who has a specific interest in chemotherapy for bone sarcoma, nominated by the cancer network clinical director and approved by the sarcoma MDT lead clinician
- d) offer all patients with bone sarcomas entry into the relevant clinical trials
- e) provide facilities for long-term follow-up for late effects of chemotherapy
- f) be guided by the bone sarcoma MDT on the treatment regimen
- g) identify an oncologist to be a member of the extended bone sarcoma MDT.

The provider of curative radiotherapy services should:

- a) provide the facilities for radiotherapy as described in the 'Manual for cancer services 2004'²²
- b) be **either**
 - at a radiotherapy centre for children and young people that meets the criteria in the NICE guidance on 'Improving outcomes in children and young people with cancer'²³ and that has a formal relationship with a bone sarcoma MDT
 - or**
 - at a cancer centre that has a formal relationship with a bone sarcoma MDT
- c) have a clinical oncologist who has a specific interest in radiation therapy for bone sarcoma, nominated by the cancer network clinical director and approved by the sarcoma MDT lead clinician
- d) be guided by the bone sarcoma MDT on the treatment regimen
- e) identify an oncologist to be a member of the extended bone sarcoma MDT.

²² Department of Health (2004) *Manual for Cancer Services*. Available from: www.dh.gov.uk (accessed 3 January 2006).

²³ National Institute for Health and Clinical Excellence (2005) *Improving outcomes in children and young people with cancer. NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

Palliation

The preferred provider for palliative radiotherapy and chemotherapy services should be decided by the sarcoma MDT in conjunction with the patient and agreed with local radiotherapy and chemotherapy providers.

*Improving Outcomes for
People with Sarcoma*

*Improving treatment:
bone sarcomas*

B. Anticipated benefits

Surgery will be performed by a surgeon with a special interest and training in bone sarcoma surgery.

A skilled team including nursing staff, pharmacists, radiographers, physiotherapists, occupational therapists and social workers with training and experience in meeting the needs of patients with sarcoma is available.

Formalising the pathway of care between the provider of surgical services and the provider of oncology services will improve the patient experience, survival and functional recovery.

Management of all patients with sarcoma by specialist MDTs including non-surgical oncology services will maximise the opportunities for recruitment into clinical trials.

6

C. Evidence

Specialist centres

A UK cohort study of patterns of care and survival in patients younger than 40 years with bone sarcoma reported that patients with Ewing's sarcoma or osteosarcoma initially treated at specialist centres have better overall survival than those treated elsewhere.

An observational study of Swedish patients with pelvic or axial chondrosarcoma reported better overall survival in patients treated at a specialist centre, when case mix was adjusted for. This study also reported that adequate surgical margins were more likely, and local recurrence was less likely, when initial surgery was performed in a specialist centre.

An Australian observational study of patients with musculoskeletal tumours reported that patients initially treated at a specialist centre were more likely to receive a complete surgical removal of their tumour.

Protocol-based care and clinical trials

The improvement in the survival of children with osteosarcoma or Ewing's sarcoma, over the period spanning the early 1970s to the mid-1980s, occurred at a time of increasing treatment in specialist centres using up-to-date protocols from clinical trials. Evidence from the more recent cohort study found that patients with Ewing's sarcoma of bone have better overall survival when treated in clinical trials, but this effect was not seen for patients with osteosarcoma.

A large observational study using data from European multicentre chemotherapy trials for patients with Ewing's sarcoma noted that, though all were treated using the same protocols, those treated in specialist paediatric oncology units had improved survival, after adjusting for other prognostic factors. The authors speculated that the improved outcome for those treated in paediatric oncology units was related to closer adherence to protocols in those units.

When treatment effects and selection bias are taken into account, however, there is little evidence of trial effects. A systematic review that compared outcomes of patients enrolled in randomised clinical trials with those receiving equivalent treatment outside the trial setting did not observe evidence for either a beneficial or harmful trial effect. A small randomised controlled trial of adjuvant chemotherapy for patients with osteosarcoma did not observe a survival difference between patients receiving chemotherapy in the trial and those treated at the same centre using the same regimen but outside the clinical trial.

D. Measurement

Structure

- membership of the sarcoma MDT as defined in Chapter 5
- availability of common protocols and referral pathways with oncology provider
- provision of and access to appropriate specialist surgical and non-surgical care
- appropriate staff levels and training in the designated chemotherapy and radiotherapy centre.

Process

- audit of compliance with nationally agreed waiting times and referral pathways
- audit of compliance with chemotherapy and radiotherapy regimens for all patients managed by a sarcoma MDT at whatever site

Outcome

- stage-specific 5-year survival and 3-year local recurrence rates for patients with bone sarcoma
- amputation rate in patients treated with curative intent
- chemotherapy-related toxic deaths
- complications of treatment, for example infection
- patient satisfaction, limb function and quality of life

E. Resource implications

By providing a more efficient and effective service and sharing good practice, there may be some resource savings to the NHS that will offset the increased resources required by the bone sarcoma treatment centres and the sarcoma MDTs.

With the exception of chemotherapy and radiotherapy, arrangements for the care of patients with bone sarcomas in England are commissioned and funded by NSCAG. In Wales, these responsibilities are held by Health Commission Wales. It is not anticipated that additional equipment or facilities will be required for the treatment of patients with bone sarcoma with regard to surgery, chemotherapy or radiotherapy.

Improving treatment: soft tissue sarcomas

Soft tissue sarcomas can arise in a variety of sites and are usually treated by a combination of surgery, chemotherapy and radiotherapy. The most common soft tissue sarcomas can be subdivided as follows:

- limb, limb girdle and truncal soft tissue sarcomas
- retroperitoneal and pelvic soft tissue sarcomas
- soft tissue sarcomas requiring shared management.

Soft tissue sarcomas can occasionally arise in other sites including the viscera and central nervous system.

Limb, limb girdle and truncal soft tissue sarcomas

The most common sites for soft tissue sarcomas are the limb, limb girdle and trunk, and these make up 60% of all cases in adults. Patients usually present with painless lumps. These tumours are currently managed by both general and orthopaedic surgeons, and where there are specialist surgeons these may have had either general or orthopaedic training. Many of these patients are currently not managed by a sarcoma MDT.

Fibromatosis is a benign but infiltrative and destructive condition that simulates soft tissue sarcoma in its physical signs and site of origin, and often in its rate of growth. Histological differentiation is crucial. Treatment is multimodal and this rare condition is within the remit of a sarcoma MDT.

A. Recommendations

Treatment recommendations (surgery, chemotherapy, radiotherapy) for all patients with limb, limb girdle and truncal soft tissue sarcoma should be decided by a properly constituted sarcoma MDT (see Chapter 5).

Patients with fibromatosis or other soft tissue tumours of borderline malignancy should be referred to a sarcoma MDT for diagnosis and management.

Surgery

All patients with limb, limb girdle and truncal soft tissue sarcoma should undergo definitive surgical resection at a soft tissue sarcoma treatment centre.

Chemotherapy and radiotherapy

There should be a formal relationship between the soft tissue sarcoma MDT and the provider of non-surgical oncology services that is characterised by common protocols, good communication, and well-defined referral pathways. This relationship should be defined in writing and approved by the cancer network director and the lead clinician in the soft tissue sarcoma MDT. Audits of compliance with these protocols will need to be demonstrated.

The provider of chemotherapy and radiotherapy services should:

- a) provide the facilities for intensive inpatient chemotherapy and radiotherapy as described in the 'Manual for cancer services', 2004²⁴.
- b) be **either**
 - at a soft tissue sarcoma treatment centre

²⁴ Department of Health (2004) *Manual for Cancer Services*. Available from: www.dh.gov.uk (accessed 3 January 2006).

or

- at a centre with a nominated medical and/or clinical oncologist who is a member of an extended sarcoma MDT (as defined in Chapter 5) and who agrees to give curative and palliative treatments (chemotherapy or radiotherapy) according to protocols defined by the sarcoma MDT. These oncologists should be nominated by the cancer network clinical director and approved by the lead clinician on the sarcoma MDT

or

- at a principal treatment centre for children or young people as described in the NICE guidance on 'Improving outcomes in children and young people with cancer'²⁵

- c) offer all patients with soft tissue sarcomas entry into the relevant clinical trials.

The sarcoma MDT should recommend the treatment regimen. All cancer networks should **either**

- host a sarcoma MDT

or

- decide to use the services of a nearby sarcoma MDT to provide all treatment facilities

or

- have a nominated medical and/or clinical oncologist who is a member of the extended sarcoma MDT (as defined in Chapter 5) and who agrees to give curative and palliative treatments (chemotherapy or radiotherapy) according to protocols defined by the sarcoma MDT. These oncologists should be nominated by the cancer network clinical director and approved by the lead clinician on the sarcoma MDT.

B. Anticipated benefits

Surgery will be performed by a surgeon with experience in soft tissue sarcoma surgery.

²⁵ National Institute for Health and Clinical Excellence (2005) Improving outcomes in children and young people with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

The patient will be treated by an oncologist who is part of a sarcoma MDT and at a centre where there is familiarity with the chemotherapy regimens and radiotherapy techniques for the management of patients with sarcoma. This will lead to consistency of care and improved outcomes.

A skilled team including nursing staff, pharmacists, radiographers, physiotherapists, occupational therapists and social workers with training and experience in meeting the needs of patients with sarcoma is available.

Outcomes and results can be recorded and expertise gained by the treatment team.

C. Evidence

A recent UK study, which adjusted for case mix, found that patients with soft tissue sarcomas of the limb, limb girdle or trunk treated at a specialist centre had better overall survival than those treated at district general hospitals.

Three studies reported survival comparisons that were not adjusted for case mix. In a population-based Canadian study, patients with soft tissue sarcoma of the limb referred to a multidisciplinary cancer centre within 3 months of diagnosis had improved overall survival and reduced risk of amputation. A Swedish study noted that patients with limb or trunk soft tissue sarcoma who were referred to a specialist centre before surgery had improved disease-free survival but not better overall survival compared with those referred to the centre following initial surgery elsewhere. A UK study observed better overall survival for children with rhabdomyosarcoma (at any anatomical site) treated in paediatric oncology centres, compared with those treated in other hospitals during the period 1977–84.

Five observational studies, one from the UK, one from France and three from Sweden, compared the surgical margins of patients with limb or truncal soft tissue sarcoma treated at specialist and non-specialist centres. None of the comparisons were case-mix adjusted. Four of the studies found adequate surgical margins were more likely for patients treated at specialist centres. The UK study did not observe a difference between the adequacy of surgical margins at specialist and non-specialist centres. There was consistent evidence, from three of the studies, that local recurrence was less likely when the initial surgery was performed at a specialist treatment centre.

Some of the above studies compared the outcomes of patients treated by a specialist sarcoma MDT and those treated by other hospitals (see Chapter 5). There was consistent evidence in favour of management by specialist sarcoma MDTs.

D. Measurement

Structure

- membership of the sarcoma MDT as defined in Chapter 5
- availability of common protocols and referral pathways with oncology providers
- provision of and access to appropriate specialist surgical and non-surgical care
- appropriate staff levels and training in the designated chemotherapy and radiotherapy centres

Process

- audit of compliance with nationally agreed waiting times and referral pathways
- audit of compliance with chemotherapy and radiotherapy regimens for all patients managed by a sarcoma MDT at whatever location

Outcome

- stage-specific 5-year survival, local control and complication rates
- patient limb function and quality of life
- patient satisfaction

E. Resource implications

The opportunity costs associated with minimum staffing levels for a sarcoma treatment centre to provide a safe and sustainable service are detailed in Chapter 5.

It is not anticipated that additional equipment or facilities will be required for the treatment of patients with soft tissue sarcoma with regard to surgery, chemotherapy or radiotherapy. However, this may require further investigation at a local level.

*Improving Outcomes for
People with Sarcoma*

*Improving treatment: soft
tissue sarcomas*

Retroperitoneal and pelvic soft tissue sarcomas

Retroperitoneal and pelvic soft tissue sarcomas pose particular challenges in treatment. They are frequently diagnosed late, and total excision with clear histological margins is rarely possible.

Radiotherapy is difficult because the tumour volume is often large and the surrounding organs (especially the small bowel, kidney, liver and spinal cord) are at risk of damage by high doses of radiotherapy.

Late diagnosis is common, often following laparotomy. The best outcomes are achieved following treatment at specialist centres where experienced surgeons and oncologists treat the patient. A high level of awareness of the possible diagnosis is required and biopsy is best avoided.

GIST and uterine sarcomas are not included in this section because their management also needs to be considered by specific treatment teams.

A. Recommendations

Patients with retroperitoneal and pelvic soft tissue sarcoma should be referred to a sarcoma treatment centre where there is a core member of the team with special expertise in managing these tumours.

NSCAG should consider commissioning designated centres for the management of retroperitoneal and pelvic soft tissue sarcomas.

B. Anticipated benefits

Further assessment and the treatment plan will be determined by a specialist MDT.

Surgery will be performed by a surgeon with a special interest in these tumours.

A skilled team including nursing staff, pharmacists, radiographers, physiotherapists, occupational therapists and social workers with training and experience in meeting the needs of patients with sarcoma is available.

When the final histology is available, cases will be reviewed by a specialist sarcoma MDT for consideration of any appropriate adjuvant therapy or entry into randomised controlled trials.

C. Evidence

The largest UK case series of patients with retroperitoneal soft tissue sarcomas describes 119 patients referred to a specialist sarcoma treatment unit between 1990 and 1995. The observation that 55% of these patients had received surgery before referral to the specialist unit suggests that many patients are being treated outside specialist centres. The lack of population-based studies in the UK, however, prevents comparisons between outcomes in different treatment settings. A Dutch observational study reported better overall survival for people with retroperitoneal sarcoma treated at a specialist tertiary referral centre compared to those treated elsewhere. Complete surgical resection was also more likely for those treated at the specialist centre. An earlier study by the same group compared the outcomes of those managed by hospitals treating more than one patient a year on average with those managed at hospitals treating fewer patients. Complete surgical resection was more likely in the higher-volume hospitals, but overall survival was not related to case volume.

In 25 institutional case series of patients with retroperitoneal soft tissue sarcoma published since 1990, hospitals admitted between 2 and 42 patients for treatment per year on average. Patients tended to present with large tumours, median size ranged from 10 to 18 cm, which were predominantly high grade. Reports of 5-year overall survival varied between 19% and 63% for patients with localised primary disease, and was between 19% and 54% when patients with recurrent disease were included. Between 40% and 96% of patients in each hospital received macroscopic surgical clearance of their tumour. The rate of surgical resection with clear microscopic margins, where reported, was considerably lower. Peri-operative mortality, where reported, ranged from 0% to 9%.

Statistical meta-analysis of patient outcomes by institutional case volume was not conducted because of important differences between the patient populations of the individual studies. Because of the rarity of retroperitoneal sarcoma, case series even from large institutions often span decades, to capture sufficient numbers for analysis. It is difficult to interpret historical improvements and institutional differences in patient outcomes because of changes in patient management practices and technologies over this time.

D. Measurement

Structure

- evidence that there is a surgeon with specific expertise in these tumours who is a core member of that MDT

Process

- evidence of participation by individual specialists at MDT meetings
- proportion of patients with these tumours referred to specialist sarcoma MDT with a specialist surgeon

Outcome

- stage-specific 5-year survival, local control and complication rates
- patient quality of life
- patient satisfaction

E. Resource implications

The opportunity costs associated with minimum staffing levels for a sarcoma treatment centre to provide a safe and sustainable service are detailed in Chapter 5.

The recommendation that NSCAG consider commissioning a number of designated centres for managing this group of patients will have resource implications that have not been considered in this review.

Soft tissue sarcomas requiring shared management

Soft tissue sarcomas can occur at a wide variety of sites in the body. There are a number of these that may present to and need management by other site-specific cancer teams. These sarcomas include:

- gynaecological sarcomas
- head and neck sarcomas
- chest wall/intrathoracic sarcomas
- skin sarcomas
- central nervous system sarcomas
- gastro-intestinal stromal tumours (GIST)
- adult-type soft tissue sarcomas arising in children.

Uterine sarcomas usually present with a pelvic mass which is sometimes asymptomatic. Often a diagnosis of sarcoma is only made postoperatively after a hysterectomy. They represent 4% of uterine malignancies – this equates to approximately 250 women with this type of tumour per year in England and Wales.

Head and neck sarcomas represent between 150 and 200 cases per year in England and Wales and between 10% and 15% of all sarcomas. No one centre is likely to have extensive experience of their treatment. There are at least 58 head and neck MDTs in England and Wales, and each of these MDTs will only see a few sarcomas per year. Frequently, there will be difficulties in making a diagnosis and patients may be treated inappropriately because of a lack of expertise about the management of this rare group of tumours. However, the skills required for the management of these head and neck sarcomas are usually similar to those required for the management of head and neck cancer, and close cooperation between the head and neck and sarcoma MDT is essential. The most crucial area where errors may lead to inappropriate management is in the histopathological assessment of the head and neck sarcoma.

The management of chest wall, intrathoracic sarcomas and pulmonary metastases requires a combination of skills available from a sarcoma MDT and a thoracic surgeon, often combined with plastic surgical reconstructive skills.

Skin sarcomas are common, and are sometimes dealt with by a skin MDT and sometimes by a sarcoma MDT. In general the larger and deeper the sarcoma the more likely it is that the patient will need to be referred to a sarcoma MDT. The care of patients with skin sarcomas is covered in more detail in the NICE guidance on 'Improving outcomes for people with skin tumours including melanoma'.²⁶

Central nervous system sarcomas are rare and will generally be managed by a neurosurgical MDT.

GISTs are the most common mesenchymal tumour to arise in the GI tract. Symptoms are often non-specific, including nausea, vomiting, diarrhoea, pain and blood loss, but are often indicative of upper GI pathology requiring urgent investigation. Although CT scanning is the standard staging investigation, positron emission tomography (PET) scans are also effective and may show up unsuspected metastatic disease. They may also demonstrate whether a patient is responding to imatinib within a few days of starting treatment. The primary treatment is surgery with wide local excision but, unlike in patients with GI carcinomas, it is not necessary to carry out routine lymph node dissection. Imatinib is the treatment of choice for patients with unresectable or metastatic GIST and it has transformed the outlook for these patients with the prospect of prolonged remission for many. Guidance on the use of imatinib in GIST was issued by NICE in 2004.²⁷ This is a rapidly changing field. Novel agents for treating GIST are likely to be available in the near future. This emphasises the importance of continued clinical research.

Adult-type soft tissue sarcomas of various types can occur in children and young people and can arise in a variety of sites in the body. Many will require chemotherapy and site-specific surgery.

A. Recommendations

The care of patients with soft tissue sarcomas requiring shared management should be managed by the appropriate site-specific MDT, the MDT for children or the MDT for young people in conjunction with a sarcoma MDT.

²⁶ National Institute for Health and Clinical Excellence (2006) Improving outcomes for people with skin tumours including melanoma. *Nice cancer service guidance* Available from: www.nice.org.uk/csgstim

²⁷ National Institute for Clinical Excellence (2004) Imatinib for the treatment of unresectable and/or metastatic gastro-intestinal stromal tumours. *NICE technology appraisal guidance* no. 86. London: National Institute for Clinical Excellence. Available from: www.nice.org.uk/TA086

The site-specific MDT has primary responsibility to liaise with the sarcoma MDT to discuss the management of each patient. Specified care plans, taking into account currently available clinical trials, should be used. It should be made clear to patients who their key worker is.

Site-specific and sarcoma MDTs need to ensure that clear pathways exist between the two MDTs, to have common treatment pathways and to clarify under what circumstances patient care should be transferred from one team to the other.

The medical management of patients with GIST should be supervised by cancer specialists with experience in the management of patients with GIST.

Clinical trials are needed for the full evaluation of imatinib, other novel agents and the role of PET scanning in GIST.

Dietetic support should be available for patients who have undergone major abdominal surgery (see the NICE guidance on 'Nutritional support in adults'²⁸).

Surgery for non-rhabdomyosarcoma soft tissue sarcomas in teenagers and young adults should only be undertaken by a surgeon with appropriate expertise, and in age-appropriate facilities, after review at a designated sarcoma MDT.

B. Anticipated benefits

Close collaboration between site-specific and sarcoma MDTs will ensure that all patients have access to appropriate expertise and advice. This should lead to better coordinated and specialist care and improved outcomes in terms of both survival and local control.

There should be increased entry into relevant clinical trials.

²⁸ National Institute for Health and Clinical Excellence (2006) Nutrition support in adults: oral nutrition support, enteral tube feeding and parenteral nutrition. *NICE clinical guideline* no. 32. Available from: www.nice.org.uk/CG032

Shared management between site-specific and sarcoma MDTs

Evidence for the organisation of care of patients according to the anatomic site of their cancer is reviewed in the NICE (and previously the NHS Executive) cancer service guidance series. There is consistent evidence that management by an appropriate site-specific specialist MDT is associated with improved patient outcomes. No studies about the collaboration of sarcoma MDTs with site-specific MDTs were found; however, expert opinion held that treatment recommendations for these rare tumours require specialist knowledge.

Imatinib

NICE technology appraisal guidance no. 86: 'Imatinib for the treatment of unresectable and/or metastatic gastrointestinal stromal tumours' recommends the use of imatinib as first-line management of patients with unresectable or metastatic GIST.²⁹ Evidence for the clinical effectiveness of imatinib was derived from six uncontrolled clinical trials, one case series and eight case reports in which patients with advanced GIST treated with imatinib showed improved survival when compared to historical controls.

The technology appraisal guidance states that the use of imatinib should be supervised by cancer specialists with experience in the management of patients with unresectable and/or metastatic GISTs.

GIST and positron emission tomography

Evidence for the use of FDG-PET for the detection of hepatic metastases from gastrointestinal cancers is considered in the assessment report accompanying NICE technology appraisal guidance no. 86: 'Imatinib for the treatment of unresectable and/or metastatic gastrointestinal stromal tumours'. A meta-analysis of non-invasive imaging methods found FDG-PET to be more sensitive than CT, MRI and ultrasound (US) methods, with equivalent specificity.

Evidence from five observational studies suggests that FDG-PET is a more sensitive indicator of early response to imatinib therapy than CT, in patients whose GISTs are measurable using FDG-PET.

²⁹ National Institute for Clinical Excellence (2004) Imatinib for the treatment of unresectable and/or metastatic gastro-intestinal stromal tumours. *NICE technology appraisal guidance* no. 86. London: National Institute for Clinical Excellence. Available from: www.nice.org.uk/TA086

D. Measurement

Structure

- referral protocols between the sarcoma MDT and the appropriate site-specific specialist MDT, which clearly define the arrangements for shared discussion and management
- appropriate specialist surgical expertise
- specialist pathology review for patients diagnosed with GIST
- provision of imatinib according to protocol

Process

- proportion of patients with these tumours whose management has been discussed at the sarcoma and/or site-specific MDTs
- audit of care and referral pathways
- proportion of patients with GIST receiving imatinib

Outcome

- patient satisfaction
- stage-specific 5-year survival, local control and complication rates
- patient quality of life

E. Resource implications

The opportunity costs associated with minimum staffing levels for a sarcoma treatment centre to provide a safe and sustainable service are detailed in Chapter 5.

Supportive and palliative care

The NICE guidance on 'Improving supportive and palliative care for adults with cancer' was published in March 2004.³⁰ This chapter complements the guidance given, with specific reference to patients with sarcomas.

The 5-year survival of patients diagnosed with sarcoma is approximately 50%, and healthcare professionals need to work together and with the patient to provide coordinated and supportive care early on in the patient's cancer journey. Patients and their carers often need a variety of support, including information on managing symptoms, and help with accessing social care and benefits. Many patients also have specific needs for orthoses and prosthetic limbs, and for a wide spectrum of rehabilitation services. Support for patients with sarcoma is a normal part of the sarcoma MDT's role. For those patients requiring palliative care, it is essential that the palliative care team in the hospital is involved early and liaises directly with the community services.

This chapter describes four key components of care:

- the key worker
- physiotherapy, occupational therapy and rehabilitation
- orthotic and prosthetic appliance provision
- specialist palliative care.

³⁰ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

The key worker

Key workers are individuals (usually specialist nurses or allied health professionals) who have an in-depth/specialist knowledge of sarcomas and their treatment and who can act as advocates of the patient, facilitating the coordination of the diagnostic and treatment pathway, providing continuity, and ensuring the patient knows how to access information and advice. They are core members of the MDT and are involved prominently and personally in the patients' overall care. They liaise with primary care teams, especially GPs, other relevant health and social care teams, psychological support teams and professionals in the community, including palliative care when necessary.

Sometimes, such as in long-term follow-up, the key worker role may be undertaken by other staff, for example a primary care team member, paediatric oncologist or other specialist, as appropriate to the care of the patient at that time.

A. Recommendations

All patients managed by a sarcoma MDT should be allocated a key worker. Patients should be provided with their key worker's name and contact details.

B. Anticipated benefits

The patient will have an identified contact for help and support at all times during their cancer journey.

C. Evidence

The NICE guidance on 'Improving supportive and palliative care for adults with cancer' considered interventions designed to improve the coordination of care.³¹ Two randomised controlled trials examined the coordination of palliative care by a hospital or community-based nurse acting as the patient's key worker. Synchronisation of care by the key worker was associated with improved patient quality of life, fewer days spent in hospital, and fewer home visits by healthcare professionals.

³¹ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

D. Measurement

*Improving Outcomes for
People with Sarcoma*

Structure

*Supportive and palliative
care*

- identification of a key worker as part of the sarcoma MDT

Process

- provision of a key worker to each patient for all stages of their treatment and care

Outcome

- improved coordination of care
- patient and carer satisfaction with the continuity of care

E. Resource implications

The resource implications of each patient having a designated key worker have been included in the cost calculated for staffing a sarcoma treatment centre (Chapter 5). The cost is based on the key worker role being undertaken by a sarcoma CNS, but in practice this role may be undertaken by any of the sarcoma MDT members.

Physiotherapy, occupational therapy and rehabilitation

Sarcoma and its treatment can have a major effect on the quality of patients' lives. Its treatment may involve an endoprosthesis (joint and bone) replacement, amputation or tumour dissection, coupled with chemotherapy and/or radiotherapy. Rehabilitation of patients with sarcoma, especially teenagers and young adults, is highly specialised. The role of the specialised allied health professionals (AHPs) on the extended MDT enables rehabilitation to be provided in a timely and coordinated way. A range of AHPs may be required at different stages in the patient's pathway, and at a range of locations. Access to these services should be coordinated by the sarcoma MDT.

Post-treatment rehabilitation helps the patient maximise the benefits of treatment, and aims to improve physical, social and emotional outcomes both during and following treatment.

Some patients will require specialist equipment such as compression hosiery, orthoses and environmental adaptations.

AHPs or key workers with appropriate experience and training can be helpful in managing problems during all stages of treatment, including side effects of chemotherapy and problems with nutrition, particularly in patients with GIST.

A. Recommendations

A specialist sarcoma physiotherapist and other specialised AHPs should be members of the extended sarcoma MDT (see Chapter 5).

Ongoing rehabilitation and supportive care should be provided locally wherever possible. This should be coordinated by the therapist in liaison with the key worker.

B. Anticipated benefits

Patients with sarcoma would receive care from trained staff familiar with their condition.

Rehabilitation would be coordinated, promoting a seamless service with the aim of achieving maximum function for those patients undergoing limb salvage or amputation.

C. Evidence

The NICE guidance on 'Improving supportive and palliative care for adults with cancer'³² reviews the evidence for the structure of effective rehabilitation services for adults with cancer.

Sarcoma-specific evidence was limited to unsystematic reviews and case reports of the rehabilitation process. No studies of the effectiveness of rehabilitation for patients with sarcoma were found.

Two review papers stressed the importance of an experienced physiotherapist, trained in the post-treatment support of people with sarcoma, in helping patients attain the best possible function. One case series described rehabilitation needs following limb-sparing surgery for osteosarcoma or Ewing's sarcoma. This study stated (without evidence) that the function of the patient's affected limb following surgery was related to adherence to a physiotherapy programme. Another case report discussed the usefulness of a written plan during the rehabilitation of a young patient with Ewing's sarcoma.

³² National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

Evidence on the rehabilitation of children and young adults with cancer is reviewed in the NICE guidance on 'Improving outcomes in children and young people with cancer'.³³ The review, limited to observational studies and expert opinion, concluded that the provision of a range of properly trained AHPs is essential during the rehabilitation of young people with cancer.

The NICE guidance on 'Improving supportive and palliative care for adults with cancer'³⁴ considers the evidence for the recommendation that AHPs should be part of cancer-specific MDTs. Evidence was limited to professional guidance for AHPs, which stressed the importance of multidisciplinary teamwork.

D. Measurement

Structure

- provision of adequately trained specialist AHPs, such as physiotherapists and occupational therapists as part of the extended sarcoma MDT
- evidence of established referral protocols
- provision of necessary equipment

Process

- evidence that patients receive adequate input from specialist therapists
- evidence of adequate arrangements for long-term care provision

Outcome

- patient satisfaction
- quality of life and functional status

³³ National Institute for Health and Clinical Excellence (2005) Improving outcomes in children and young people with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

³⁴ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

E. Resource implications

The resource implications of designated key workers and sarcoma physiotherapists are reported in Chapter 5. Any additional staffing to ensure that minimum staffing levels are met will need to be investigated by local commissioners.

Orthotic and prosthetic appliance provision

Many patients with sarcoma, especially those who have had surgery for a limb tumour, may need to use orthoses because of reduced function. Some patients need to have a limb amputation and they will require lifelong access to specialist rehabilitation services, specifically for the supply and ongoing fitting of prosthetic appliances. Most patients who require an amputation have a bone sarcoma. The majority are young (median age 21 years) and active prior to the diagnosis of sarcoma. The aspirations of this group include the return of normal mobility, and a return to work and leisure pursuits.

Current prosthetic provision in the UK is variable – a survey undertaken by the Audit Commission in 2000 found 25% of patients fitted with prosthetic limbs found them unusable. There is evidence that non-use of prosthetics is related to the weight of the appliance, its unwearability with certain clothing, and its appearance.

Presently Disablement Service Centres (DSCs) provide prostheses in the UK. There are 44 DSCs in the UK of which 14 match the template for specialist Prosthetic and Amputee Rehabilitation Centres (PARC) proposed by the British Society of Rehabilitation Medicine (BSRM) in 2003. These centres have the necessary expertise and facilities to cope with all the intricacies of comprehensive prosthetic/amputee rehabilitation.

A. Recommendations

Rapid, easy access should be provided to appropriate orthotic and prosthetic services.

The sarcoma MDT should establish formal links to a centre(s) matching the PARC template, and should refer patients for pre-amputation assessment.

Special activity limbs should be provided where appropriate and proven technological improvements should be made available.

B. Anticipated benefits

Improved functioning and quality of life

Improved integration into society.

C. Evidence

Satisfaction with prosthetics services

An Audit Commission report in 2000 identified user concerns with aspects of the prosthetics service in the UK, especially with regard to information provided to patients. The same report also found that approximately 25% of patients fitted with prosthetic limbs found them unusable for reasons of discomfort, pain, poor fit and appearance.

In a 2002 update to the original Audit Commission report, some improvements and examples of innovative practice were noted. An observational study, reporting high satisfaction levels in users of three UK DSCs, suggests that examples of good service provision exist.

Specialised rehabilitation service

Expert opinion held that many DSCs currently lack the expertise to deal with all aspects of the orthotic and prosthetic rehabilitation of patients with sarcoma. It was thought that sufficient expertise should, however, be available in those DSCs meeting the specifications of a Tertiary Referral PARC as defined in the BSRM standards and guidelines for amputee and prosthetic rehabilitation.

Prosthetics for leisure activities

A cohort study, reporting the incidence and aetiology of limb amputation in the UK (2003–2004), found that people who lose a limb as the result of a primary tumour tend to be younger than other amputees. Consequently the rehabilitation aspirations of this group may exceed basic mobility and include return to work and leisure pursuits.

D. Measurement

Structure

- provision of adequately trained specialist orthotists and prosthetists
- provision of special activity limbs

Process

- operational policies both pre- and postoperative between sarcoma treatment centres and PARCs
- evidence of long-term care provision

Outcome

- patient and carer satisfaction with orthotic and prosthetic appliance provision.

E. Resource implications

It is not known how many people who have sarcoma-related amputations will require an activity limb, or indeed how many already have one. The costs are presented for two scenarios. If all those patients with sarcoma (53) in England and Wales who have transfemoral amputations require a water activity limb, the cost would be £480,000. If members of the same group require a limb with a computerised knee and cosmesis, the cost would be £1,200,000. The cost impact of this aspect of the guidance will be determined by patient choice.

Specialist palliative care

Palliative care is essentially a community service and needs to be provided locally for patients with sarcoma as required. There is, however, also a need for specialist palliative care input for some patients at the time of MDT discussion, and there should be access to specialist palliative care teams within the hospital and community.

A. Recommendations

A member of the specialist palliative care team should be a member of the core sarcoma MDT.

Key workers should have a major role in liaising with palliative care and support services such as hospice and Macmillan services.

Commissioners should ensure that patients with sarcoma have easy and timely access to appropriate palliative and specialist pain management services (see the NICE guidance on 'Improving supportive and palliative care for adults with cancer'³⁵).

³⁵ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

B. Anticipated benefits

Improved integration of palliative care services throughout the course of the illness will enhance quality of life for both patients and their carers.

Provision of patient-centred, holistic care and clear and timely information will help patients to cope with their disease, enhance satisfaction with services and reduce complaints.

Integrated care is particularly important at the end of life, and the contribution of palliative care specialists will help to create a more appropriate balance between efforts to preserve life and the need for comfort, peace and support for close family members when it becomes clear that death is inevitable.

C. Evidence

Palliative care specialists with an interest in sarcoma

No evidence was identified on the effectiveness of specialist palliative care teams with an interest in sarcoma. The NICE guidance on ‘Improving supportive and palliative care for adults with cancer’³⁶ reviews evidence for the configuration of palliative care services.

Evidence from seven systematic reviews supports the effectiveness of specialist palliative care teams for the control of pain and symptoms of people with cancer. Patients cared for by specialist teams were more satisfied than those cared for elsewhere.

Shared specialist palliative care between the MDT and local services

Evidence from systematic reviews suggests that specialist palliative care delivered at a patient’s home or in a hospice can be as effective as conventional hospital-based care in the control of pain and symptoms and in terms of patient satisfaction.

The composition of the specialist palliative care team

There was insufficient evidence to recommend the ideal structure, but patient outcomes tended to be better with specialist palliative care teams made up of multidisciplinary trained staff.

³⁶ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

Coordination between hospital and community-based teams

Two randomised controlled trials reported that employment of a nurse coordinator, who provided a link between patients and the health services, reduced the number of days spent in hospital by the patient and the number of home visits by the community care team.

D. Measurement

Structure

- specialist palliative care teams to support patients at home or in hospices
- telephone support, advice and information services for patients and their carers
- bereavement counselling for family members and carers as appropriate

Process

- attendance of the palliative care specialist at the sarcoma MDT
- evidence that providers elicit information about patient preferences relating to place of death and their views about medical intervention in the terminal phases of illness
- regular systematic psychological assessment at key points, and access to appropriate psychological support as recommended in NICE guidance on 'Improving supportive and palliative care for adults with cancer'³⁷

Outcome

- patients' experience of pain and satisfaction with pain control during treatment
- symptom control and quality of life
- patient and carer satisfaction with the services provided during the patient's final month of life

³⁷ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

E. Resource implications

*Improving Outcomes for
People with Sarcoma*

*Supportive and palliative
care*

The costs of providing specialist palliative care are presented in Chapter 5. It will be for local commissioners to investigate any additional healthcare professionals who may be required on a centre basis. It is likely that patients will access palliative care and support more locally instead of at a sarcoma treatment centre, and the resource implications for this are included in the NICE guidance on ‘Improving supportive and palliative care for adults with cancer’.³⁸

³⁸ National Institute for Clinical Excellence (2004) Improving supportive and palliative care for adults with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/page.aspx?o=110005

Follow-up of patients

The main aim of follow-up is to detect recurrent disease at a time when treatment can favourably influence the outcome for that patient. An additional reason is to assess and deal with any complications of treatment and to reassure patients. Local recurrence rates in the UK average 10–20% for extremity soft tissue sarcomas and around 10% for most bone sarcomas, but may reach as high as 30% for large high-grade tumours excised with a close margin, compared with 5% for low-grade tumours excised with a wide margin. Up to 50% of patients with high-grade soft tissue sarcomas will develop lung metastases but low-grade or subcutaneous tumours have a much lower risk. The value of follow-up has been questioned because 40% or more of patients with bone and extremity soft tissue sarcomas will never develop a recurrence and so will never need following up. It is also time-consuming and expensive and may produce anxiety rather than reassurance for patients.

For patients with GIST there is a much higher risk of recurrence that is likely to be intra-abdominal.

For sarcoma patients enrolled in clinical trials, there will usually be a standard follow-up regimen suggested by the trial protocol. The basic minimum follow-up would include careful clinical examination and a chest X-ray at regular intervals. The value of more sophisticated investigations remains uncertain for the detection of both local recurrence and metastatic disease, but may be specific for individual tumours (for example PET or CT for GIST).

Long-term follow-up will be needed for many patients, especially those who have received a prosthetic replacement and those who had a childhood cancer, because of the risk of late complications. Some patients have a genetic element to their disease and this may cause concern to them and their families.

A. Recommendations

Research should be commissioned to provide evidence for the follow-up protocols required for each tumour type.

Resources should be made available for regular imaging of patients at high risk of recurrence (as defined in an agreed protocol, for example the American National Comprehensive Cancer Network/American College of Radiology consensus-based guidelines).³⁹

Where appropriate, access to cancer genetic services should be offered to the patient and their family.

B. Anticipated benefits

Clearly defined protocols for follow-up will improve the consistency and equity of care for these patients and the appropriate use of resources.

Early detection of recurrent disease is of benefit.

C. Evidence

Current situation

A review article identified 11 papers in which experts recommended 26 strategies for the follow-up of extremity soft tissue sarcoma. There was consensus on the importance of routine clinical examination and chest X-ray in follow-up. There was disagreement, however, over the role of routine chest CT, and over the best method for regular imaging of the primary site.

A survey of 318 American surgeons about post-treatment follow-up protocols for extremity soft tissue sarcoma showed considerable variation in strategies. Clinical examination and chest X-ray were the most frequently performed follow-up tests. Approximately half the surgeons ordered MRI or CT imaging of the primary site in the first postoperative year. The frequency of follow-up visits was usually related to an estimated risk of recurrence, based on the time elapsed since treatment, tumour characteristics and surgical margins. A recent National Cancer Research Institute (NCRI) study in the UK produced very similar findings.

³⁹ American College of Radiology *Follow-up examinations for bone tumors, soft-tissue tumors, and suspected metastasis post therapy*. Available from: www.acr.org (accessed 3 January 2006)

The American National Comprehensive Cancer Network and the American College of Radiology have issued consensus-based guidelines for the follow-up of sarcoma,⁴⁰ which propose strategies stratified by the grade and site of the original tumour.

Effectiveness of follow-up strategies

No studies were found which compared follow-up strategies for patients with sarcoma in terms of health outcomes.

An observational study reported the effectiveness of routine follow-up for the detection of recurrence in patients with primary extremity soft tissue sarcoma at an American treatment centre. Altogether, 29/141 patients developed a local recurrence, all but one of which was discovered during physical examination. The importance of patient education in follow-up is supported by the fact that 13/29 of the local recurrences were detected either by the patient or a primary care doctor between follow-up visits.

None of the 21 patients who presented between follow-up visits with symptomatic pulmonary metastases were considered candidates for potentially curative surgical resection of their metastases. Resection of pulmonary metastases was performed for 24 of the 36 patients whose asymptomatic recurrence was discovered by surveillance chest X-ray or staging CT scan.

The effectiveness of routine follow-up testing was also considered in an observational study of 643 patients at a UK sarcoma treatment centre. For patients with soft tissue sarcoma, 15% of local recurrences were discovered at a follow-up appointment and 70% were detected by the patient between surveillance visits. For bone sarcomas, 36% of local recurrences were picked up at surveillance visits and 57% were discovered by the patient.

Evidence on the acceptability of follow-up to people with sarcoma was limited to a small cross-sectional study of 30 patients. Although patients reported anxiety before follow-up visits, 80% said that the visit itself was a positive experience.

⁴⁰ American College of Radiology *Follow-up examinations for bone tumors, soft-tissue tumors, and suspected metastasis post therapy*. Available from: www.acr.org (accessed 3 January 2006)

Late effects of treatment

Evidence from cross-sectional studies, reviewed in the NICE guidance on 'Improving outcomes in children and young people with cancer', suggests that most patients have at least one moderate to severe adverse health outcome following treatment for childhood cancer.⁴¹ A European observational study recorded late effects in the year following cessation of therapy in clinical trials for Ewing's sarcoma, osteosarcoma or soft tissue sarcoma. At this relatively early stage, cardiotoxicity was noted in 12%, ototoxicity in 7% and nephrotoxicity in 1% of patients.

D. Measurement

Structure

- nationally agreed protocols for follow-up

Process

- audit of follow-up practices and the timeliness and appropriateness of investigation

Outcome

- patient/carer satisfaction

E. Resource implications

The opportunity costs associated with minimum staffing levels to ensure that all MDTs have sufficient capacity and expertise to manage and care for long-term follow-up have been included in the MDT section (Chapter 5).

The resource implication of the recommendation that research is commissioned to provide evidence for follow-up protocols for each tumour type has not been included in this review.

It is recommended that patients are referred to cancer genetic services where appropriate. The capacity within existing cancer genetic services will require further investigation by local commissioners.

⁴¹ National Institute for Health and Clinical Excellence (2005) Improving outcomes in children and young people with cancer. *NICE cancer service guidance*. Available from: www.nice.org.uk/CSGCYP

Improving knowledge

Sarcomas are rare tumours, and reliable information on management and outcomes is largely limited to individual units that manage the conditions.

Data collected for needs assessment and audit purposes has shown the lack of a systematic dataset across England and Wales, making meaningful comparison of clinical processes and outcomes difficult. The two audits of soft tissue sarcoma carried out in England over the last 10 years demonstrated poor compliance with agreed best practice. No single disease register exists for sarcomas in England and Wales. In Scandinavia, the existence of a multicentre sarcoma dataset has led to better monitoring of outcomes, and systematic improvements in referral, diagnosis and treatment.

Implementation of the nationally agreed dataset for sarcoma as a subset of the National Cancer Dataset⁴² will enable multicentre audit to be carried out, and enable clinicians to add to the overall level of knowledge of disease management and outcome data. It is at present unclear how the data for this dataset will be collected and who will have ownership of the data.

Training and CPD are key factors in maintaining and improving standards of care. Apart from the EQA scheme for pathologists there is no current training or quality assurance for those involved in sarcoma care.

A. Recommendations

Data collection

All sarcoma MDTs should collect data on patients, tumour, treatment and outcome.

The data collected should be agreed nationally and should be based on the sarcoma subset of the National Cancer Dataset (including comorbidity data). Cancer networks should ensure that a complete dataset exists for all patients managed within their network.

⁴² National Cancer Dataset. www.icservices.nhs.uk/datasets/pages/cancer/datannuals.asp

Public health observatories or cancer registries should act as the data repository of the agreed dataset, and a lead observatory or cancer registry should be commissioned as the repository of a national dataset, which could then become a national sarcoma register.

Audit

Audit should be carried out of all elements of the referral and management pathway including standards for referral, investigation and management.

SSPs should continue to undertake the existing EQA scheme, and networks should ensure that only specialist sarcoma pathologists who comply with this scheme report on sarcomas.

Commissioners should ensure that networks and sarcoma MDTs audit the management of sarcoma on a regular basis, using the national dataset for comparison of compliance with management guidelines and outcomes. The National Clinical Audit Support Programme should be asked to provide guidance on multicentre audits.

National audits of outcome including patient satisfaction should be carried out by networks and sarcoma MDTs.

The results of audits should be widely available to clinicians within referring units, networks and the public.

Training

Commissioners should ensure that all those involved in sarcoma care remain up to date with current advances in sarcoma care, and can provide evidence of adequate, relevant CPD.

Appropriate training posts should be made available nationally to train and recruit surgeons, pathologists, radiologists and oncologists with appropriate expertise in sarcoma care.

Training should be developed and provided for all members of both the core and extended sarcoma MDTs.

The Royal Colleges of Surgeons should be invited to make recommendations about appropriate training and certification of sarcoma surgeons.

MDTs should ensure that they regularly provide updates for members of the extended MDT.

Research

Improvements in the management of sarcomas require reliable evidence that interventions are effective and that they improve outcomes for patients. There is limited evidence-based information available on many aspects of the management of sarcoma, including the optimum patient pathway and the configuration of services. It is therefore important that health service commissioners should support the well-designed clinical trials within the portfolio of the National Cancer Research Network (NCRN), which should be encouraged to investigate diagnostic pathways.

Data from the national dataset for sarcoma should be used for research purposes to enable multicentre survival studies to be carried out on a relatively large and complete population base.

Commissioners should ensure that NCRN-adopted clinical trials for patients with sarcomas are supported locally.

All sarcoma MDTs should aim to maximise entry into trials and should work with the local NCRN to ensure this happens. They should have a nominated research lead.

The possibility of entry into an appropriate trial should be discussed with every patient who fits the inclusion criteria. Such patients should be given accurate and accessible information to inform their decision about whether to participate in the trial.

Trials of treatment for sarcoma should be designed with outcome measures that reflect quality of life, including the use of limb prostheses in bone sarcoma (assessed by patients, not just clinicians), as well as survival time and clinical measures with prognostic significance.

Patients who are not involved in a clinical trial should be treated according to local clinical guidelines based on research evidence.

B. Anticipated benefits

Data collection and audit

In the case of rare cancers such as sarcoma, it is only possible to audit the compliance with management guidelines by pooling data. The use of a large standard dataset will facilitate multicentre audit.

Entering individual patient data into a national database will encourage sarcoma MDTs to compare their own performance against that of their peers, and lead to improvements in compliance with the guidance.

Training

Ensuring that all healthcare professionals managing patients with sarcoma are appropriately trained and are kept up to date with recent developments will improve and maintain the quality and effectiveness of the service.

Training posts will encourage new enthusiasm for sarcoma care across all the treating specialities.

Research

Reliable information on the effectiveness of clinical interventions can only be obtained from large, well-designed trials. Wider participation in such trials will increase the evidence base.

C. Evidence

Data collection

The rarity of sarcomas means that data need to be pooled before analysis. The Scandinavian Sarcoma Group central register of soft tissue and bone tumours, for example, collects information from all treatment centres in Finland, Norway and Sweden. This register allows evolving treatment patterns and patient outcomes to be monitored, and enables regular audit of patient management against recommendations.

Evidence from three observational studies, including one from the UK, suggests that central review of histopathology by specialist tumour registry pathologists improves diagnostic accuracy. In the UK study, clinically important diagnostic errors were detected in 8% of the cases submitted to a bone tumour registry.

An unpublished study reported 88% concordance between new diagnoses of soft tissue sarcoma recorded in the South West of England Cancer Registry in 2003, and those identified in histopathological audit data. The authors suggested that improving the quality of data sent to cancer registries would reduce the need for a separate sarcoma register.

Clinical trials and protocol-based care

The survival of children with osteosarcoma or Ewing's sarcoma has shown great improvement over the last three decades, a time during which treatment has been increasingly given in specialist centres using protocols from clinical trials. Evidence from more recent observational studies suggests that patients with Ewing's sarcoma of the bone have better survival when treated using protocols from clinical trials. Regional audit data from the UK, however, showed that less than 50% of people with soft tissue sarcoma were enrolled in clinical trials, and no evidence was found on the effect of treatment in clinical trials on outcomes for this group of patients.

Although the evidence in sarcoma is far from definitive, treatment in accordance with local clinical guidelines (protocols) is generally associated with better outcomes in other cancers (see for example NICE guidance on 'Improving outcomes in breast cancer'⁴³). Expert opinion held that the development of local protocols demands a critical attitude towards best practice, which is likely to have a beneficial effect for patients.

D. Measurement

Data collection

Structure

- network-wide information systems that capture standard data of all patients with sarcoma according to the National Cancer Dataset
- availability of support to collect data and enable it to be shared within and between networks

Process

- evidence that all patients' data are collected in accordance with national protocols
- evidence that pathology data are collected and that the completeness of that data can be subjected to audit

⁴³ National Institute for Clinical Excellence (2002) Improving outcomes in breast cancer. *NICE cancer service guidance*. Available from www.nice.org.uk//page.aspx?o=204541

Audit

Structure

- availability of support to carry out multicentre audits
- agreements between cancer networks and sarcoma MDTs about the audit tool and frequency of multicentre audits
- support for patient groups for carrying out patient satisfaction audits on behalf of patients with sarcoma

Process

- proportion of sarcoma MDTs participating in regular audit and multisite audits
- evidence of feedback and the development of action plans to referring clinicians following audit

Clinical trials

Structure

- network-wide information systems that allow clinicians to identify trials for which specific patients might be eligible
- availability of support for clinical trials
- availability of continued support for patients who have been successfully treated with products used in clinical trials

Process

- evidence of regular discussion of participation in clinical trials at MDT meetings

Outcome

- proportion of patients with each type of sarcoma entered into trials

E. Resource implications

Data collection

The MDT section includes each MDT having a full-time equivalent (FTE) coordinator with clerical support; this provision will ensure that there is full recording of comprehensive patient data.

It is anticipated that there would need to be an additional full-time data manager or research officer post at the lead observatory or cancer registry commissioned to become holder of the national sarcoma dataset. The employment costs of a FTE data manager will be around £34,788 per year. This would vary in line with the exact requirements of the post and the experience of the data manager.

Audit

The resource implications of recommendations concerning audit have not been included in this review.

Training

The cost of four training sessions per year for staff at diagnostic clinics that are not based at a sarcoma treatment centre have been estimated to be between £1805 and £2561, dependent upon whether it is a sarcoma CNS-led clinic or a consultant-led clinic.

Additional specialist sarcoma training may be required in some centres for nursing and other healthcare professionals, such as the nursing modules offered by the University of Central England in conjunction with the Royal Orthopaedic Hospital. The costs per module are likely to be around £220.

Research

The resource implications of the research recommendations have not been formally costed. Priorities on allocating research funds are made by national, government and charitable medical research funding agencies.

Scope of the guidance

A1

1. Guidance title

Service Guidance for Improving Outcomes for People with Sarcoma

1.1 Short title

Sarcoma

2. Background

- a) The National Institute for Clinical Excellence ('NICE' or 'the Institute') has commissioned the National Collaborating Centre for Cancer to develop service guidance on sarcomas for use in the NHS in England and Wales. This follows referral of the topic by the Department of Health and Welsh Assembly Government (see page 107). The guidance will provide recommendations for service provision that are based on the best available evidence.
- b) The Institute's service guidance will support the implementation of the National Service Frameworks (NSFs) in those aspects of care where a Framework has been published. The guidance will support current national initiatives outlined in the *NHS Cancer Plan*, the Calman Hine report, the Cameron report, the *Manual of Cancer Service Standards for England* and the *All Wales Minimum Standards for Cancer Services*. Cross-references will be made to these and any other documents as appropriate.

The guidance will also refer to other NICE documents currently under development, including Referral Guidelines for Suspected Cancer, Supportive and Palliative Care for People with Cancer, Service Guidance for Improving Outcomes in Child and Adolescent Cancer, Head and Neck Cancers, Haemato-Oncology, Skin Tumours including Melanoma and Tumours of the Brain and Central Nervous System.

Cross-references will be made to these and any other documents as appropriate.

3 Clinical need for the guidance

- a) Sarcomas are a rare and heterogeneous group of tumours. They may arise in either bone or soft tissue. There are between 1500 and 2500 new cases of soft tissue and bone sarcoma and gastrointestinal stromal tumours (GIST) per year in the United Kingdom, accounting for 1% of adult malignancies and 6% of those in childhood.
- b) Soft tissue sarcomas involve the connective tissues and usually present as a swelling in any part of the body including limbs, trunk and head and neck. There are many histological types, with a range of clinical behaviours from local invasion to distant metastasis. Because these tumours are rare, the diagnosis is frequently not suspected at presentation and subsequent management can be variable. Bone sarcomas present with bone pain, often with swelling – osteosarcoma occurs most frequently, with a peak incidence in adolescence.

Treatment generally consists of surgery, combined with radiotherapy and chemotherapy. However, patients frequently require complex management and the best organisation of care has yet to be determined.

4 The guidance

- a) The guideline development process is described in detail in three booklets that are available from the NICE website (see Section 5, 'Further information'). *The Guideline Development Process – Information for Stakeholders* describes how organisations can become involved.
- b) This document is the scope. It defines exactly what this piece of service guidance will (and will not) examine, and what the developers will consider. The scope is based on the referral from the Department of Health and Welsh Assembly Government (see page 107).
- c) The areas that will be addressed by the guideline are described in the following sections.

4.1 Population

4.1.1 Groups that will be covered

- a) All patients with malignant bone tumours and those tumours of unspecified, borderline and uncertain behaviour as defined by the World Health Organization (WHO) classification (*World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of Soft Tissue and Bone*, IARC Press, Oxford, 2002, ISBN 9283224132).
- b) All patients with malignant soft tissue sarcoma and those tumours of unspecified, borderline and uncertain behaviour as defined by the WHO classification, excluding Kaposi's sarcoma because as this is included in the *Service Guidance for Skin Tumours, Including Melanoma* (www.nice.org.uk/pdf/Skin_scope.pdf).
- c) All patients with gastrointestinal stromal tumours (GIST).

4.1.2 Groups that will not be covered

Adults and children with:

- benign bone and soft tissue tumours as defined by the WHO classification
- metastases to bone and soft tissues from tumours at other primary sites.

4.2 Healthcare setting and services

- a) Primary care, including diagnosis, treatment and follow up.
- b) Secondary care, including the role of cancer networks and multidisciplinary teams (MDTs).
- c) Tertiary care in cancer centres and specialist surgical units (for example, thoracic and plastic surgery).
- d) Quaternary care in specialist regional or national units for appropriate patients (for example, bone tumour surgery).

4.3 Key areas of clinical management

The following key areas of clinical management will be included, because they have direct implications for service delivery. However, because of the potential overlap with the *Service Guidance for Child and Adolescent Cancer*, chemotherapy and support services for

children and young people in their late teens and early twenties will not be included within this guidance, but it will be cross-referenced.

a) Services for diagnosis and staging (excluding those being addressed as part of the updated referral guidelines) including:

- primary care
- surgical services in secondary care
- pathology departments
- diagnostic radiology departments.

In addition, the guidance will address the important issue of data collection and registration of sarcomas.

b) Treatment services, to include treatment in the following settings:

- surgical services – DGHs, cancer units, cancer centres, specialist surgical units and quaternary centres
- oncology services – cancer units and cancer centres.

c) Follow-up (need, frequency, type, location, and by whom) to include the surveillance of patients with conditions known to predispose to sarcoma.

d) Specific elements of supportive and palliative care that meet the particular needs of patients with bone and soft tissue sarcoma.

e) Rehabilitation and support of patients with bone and soft tissue sarcoma, including the role of specialist nurses, physiotherapy, occupational therapy and disablement services.

f) Information resources and support for patients, carers and family members.

g) Health service research and clinical trials on service delivery.

4.4 Audit support within the guidance

The guidance will include key criteria for audit, which will enable objective measurements to be made of the extent and nature of local implementation of this guidance, particularly its impact on practice and outcomes for adults with sarcoma.

4.5 Status

4.5.1 Scope

This is the final version of the scope.

4.5.2 Guidance

The development of the service guidance recommendations will begin in February 2004.

A1

5 Further information

Information on the guideline development process is provided in:

- *The Guideline Development Process – Information for the Public and the NHS*
- *The Guideline Development Process – Information for Stakeholders*
- *The Guideline Development Process – Information for National Collaborating Centres and Guideline Development Groups.*

These booklets are available as PDF files from the NICE website (www.nice.org.uk). Information of the progress of the guideline will also be available from the website.

Appendix – Referral from the Department of Health and Welsh Assembly Government

The Department of Health and Welsh Assembly Government asked the Institute:

“To prepare service guidance for the NHS in England and Wales for sarcoma. This would form part of the *Improving cancer outcomes* series with NICE expected, as previously, to involve the Department of Health and Welsh Assembly Government closely in the development of the guidance. In particular, the Department of Health and Welsh Assembly Government should be alerted at an early stage to any issues in the developing guidance, which are likely to lead to significant changes in the current service provision.”

Appendix 2

Economic implications of the guidance

A2

Executive summary

The economic consequences of the recommendations of the ‘Guidance on cancer services: improving outcomes for people with sarcoma’ in England and Wales are set out in this document. The analysis focuses on those aspects of the key recommendations that are likely to be of greatest consequence in terms of cost and this varies according to type of sarcoma. Bone sarcomas are currently treated centrally, whereas soft tissue sarcomas are treated more disparately. Moving to a more centralised service as proposed by the manual will have cost implications.

The summary of economic implications is outlined in Table A1.

There is some uncertainty around the estimates presented and there will be variation between costs for different diagnostic clinics and sarcoma treatment centres. Therefore sensitivity analyses were conducted to account for uncertainty in the estimated costs. Further assessments will be needed at cancer network level and/or NHS trust level to determine the exact cost implications. Work is currently being carried out in the NHS in England, in connection with ‘Payment by results’, to develop a better understanding of costs of treatment and care. This may help these assessments in the future.

Information from two specialist hospitals that treat patients with soft tissue sarcoma suggests that the Healthcare Resource Groups (HRGs) currently used for the funding of major surgery significantly underestimate the true costs of the procedure and inpatient care. Although these HRGs have not been used in this economic assessment, it is important that commissioners take this into account when calculating the overall costs of services.

Table A1. Summary of estimated annual economic implications

	Costs per year (£)	
	Low range	High range
Annual employment costs of a new diagnostic clinic	88,833	101,622
Average unit cost of triple assessment for patients referred to diagnostic clinics in England and Wales ^a	213.00	
Average unit cost of MRI scans needed, in addition to ultrasound, to confirm diagnosis in 30% of all patients referred to diagnostic clinics in England and Wales	223.94	
Employment costs of additional staff required at existing molecular pathology/cytogenetics laboratories	79,950	
Core employment costs of a sarcoma treatment centre	482,399	819,039
Cost of producing information leaflets (for all patients with sarcoma in England and Wales)	26,420 (year 1) 17,300 (subsequent years)	
Cost of a National Implementation Group (for England and Wales)	98,443	100,843
Cost of orthotic and prosthetic appliances (per cancer network)	5,622	28,649
Employment cost of having a lead cancer registry (for England and Wales)	34,788	

^a Based on cost estimates for triple assessment for the diagnosis of breast cancer.
MRI – magnetic resonance imaging.

Improving diagnostic services

Diagnostic clinics

The annual opportunity cost for each diagnostic clinic operating one diagnostic session per week for 45 weeks of the year, inclusive of four training sessions per year, is estimated to be between £88,833 and £101,622. The variation is dependent upon whether the clinic is lead by a doctor or a sarcoma CNS.

These costs do not take into account the cost of any new equipment, such as ultrasound, that may be needed to provide this diagnostic service. These values represent the estimated maximum cost of staffing a new diagnostic clinic.

Triple assessment

The guidance also recommends triple assessment of all patients referred to a diagnostic clinic with a suspected soft tissue sarcoma. National reference costs for triple assessment in the diagnosis of sarcoma are not available, so costs for breast triple assessment have been used to give an indication.

The average unit cost for providing breast triple assessment is £213 with an interquartile range of between £150 and £242. We estimate that there would be around 20,000 new referrals per year so the cost impact of triple assessment for all of these patients would therefore be approximately £4,260,000 (interquartile range £3,000,000 to £4,840,000).

Triple assessment for sarcoma is likely to be more expensive, as approximately 30% of the 20,000 patients referred for diagnostic tests will require a magnetic resonance imaging (MRI) scan in addition to an ultrasound, to confirm their diagnosis. The average unit cost for an MRI scan is £223.94 (interquartile range of between £184 and £465); therefore the additional cost for this patient group would be approximately £1,343,640 in England and Wales (interquartile range £1,164,000 to £2,790,000).

Cytogenetics/molecular pathology laboratories

The Guidance Development Group anticipates that additional staff will be required at the existing cytogenetic/molecular pathology facilities, to undertake the work generated as a result of implementing the guidance. It is anticipated that there would need to be an additional clinical scientist, biomedical scientist and administrative support employed at each of the laboratories. The additional employment cost per existing laboratory would be around £79,950.

Improving treatment

The opportunity costs for minimum staffing levels at a sarcoma treatment centre have been estimated. The annual employment cost of the medical, nursing and other staff caring for 100 new patients per year is estimated to be between £482,399 and £819,039 per year. The cost calculations are for members of the multidisciplinary team (MDT), ward nurses and outpatient nurses, but are not inclusive of all staff who would be involved with the patients' care. Other clinical staff and the ancillary, catering or administration workforce would be an additional cost.

There is likely to be an additional requirement for some healthcare professionals, in particular sarcoma CNS and specialist sarcoma physiotherapists. This needs to be considered by local commissioners. As with costs associated with the employment of staff at the diagnostic clinics, it needs to be emphasised that these costs represent opportunity costs because the staff involved in the treatment centres are already contracted to the NHS. However, at present the staff are employed in a variety of locations rather than in designated sarcoma treatment centres.

Patient perspective

The resource implications of providing information leaflets for sarcoma patients throughout England and Wales are expected to be £26,420 for the first year and £17,300 for subsequent years. This cost assumes the production of up to eight generic leaflets on different types of sarcoma and also the production of diagnostic clinic/sarcoma treatment centre-specific leaflets.

National Implementation Group

It is anticipated that the National Implementation Group would have a wide-ranging function including establishing an expert board to develop a comprehensive strategy to implement and monitor the guidance at all levels.

It is assumed that the National Implementation Group would comprise a full-time manager and one or two administrative support workers, and a public health doctor and specialist commissioner (both on a sessional basis). The group would establish and facilitate a board (meeting 3 or 4 times a year). The estimated resource implications would be between £98,443 and £100,843 per annum.

Orthotic and prosthetic appliance provision

It is not known how many people who have sarcoma-related amputations will require an activity limb, or indeed how many already have one. We have presented a sample of costs for patients with sarcoma who have undergone transfemoral amputations. The annual cost estimates vary from £5622 to £11,459 per network for 50% of transfemoral amputees to have a water activity limb. For all sarcoma-related transfemoral amputees to have an activity limb with computerised knee and cosmesis, the cost is estimated to be between £14,054 and £28,649 per network. The cost impact of this aspect of the guidance will vary in line with patient choice.

Improving knowledge

It is anticipated that there would need to be an additional full-time data manager or research officer post at the lead observatory or cancer registry that is commissioned to become holder of the national sarcoma dataset. The employment costs of a data manager (Agenda for Change Band 6 point 30) will be around £34,788 per year. This would vary in line with the exact requirements of the post and the experience of the data manager.

How this guidance manual was produced

This service guidance is intended to guide health organisations (strategic health authorities, primary care trusts, local health boards, cancer networks and trusts), their managers and lead clinicians in improving the effectiveness and efficiency of services for people with sarcoma. The information and recommendations in the manual are based on reviews of the best available evidence on diagnosis, treatment and service delivery. This evidence is retrieved by information specialists and assessed by researchers within the National Collaborating Centre for Cancer (NCC-C) and the recommendations are the product of extensive discussion with the Guidance Development Group (GDG). A brief overview of the development process to produce the guidance is provided below.

The first stage in the development of the guidance was the production of a scope (Appendix 1), which defined in detail the patient population, the healthcare settings and services, and key areas of clinical management that the guidance should cover. This was then subject to a 4-week consultation with registered stakeholders in line with National Institute for Health and Clinical Excellence (NICE) methodology. Following this a multidisciplinary GDG was formed comprising clinicians representing the main stakeholder organisations and representatives from relevant patient organisations and charities (Appendix 4.1). The GDG was convened by the NCC-C and chaired by Dr Joe Kearney in close association with the Clinical Lead, Mr Robert Grimer. All GDG members made and updated any declarations of interest. The Group met on a monthly basis during development of the guidance, and NCC-C staff provided methodological support and leadership for the development.

During the development phase of the guidance the GDG identified areas where there was a requirement for expert input on particular specialist topic areas. These topics were addressed by the production of a position paper by a recognised expert who had been identified via the relevant registered stakeholder organisation. All relevant expert positions papers are presented in Appendices C to D of the Evidence Review.

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The identification and retrieval of evidence to support the recommendations in the guidance manual is described in detail in the Evidence Review. Briefly, there were three stages to this process:

- Clinical question development. Members of the GDG were asked to submit clinical questions to the NCC-C on issues covered by the project scope.
- Literature searching. All clinical questions were prioritised and were subject to a systematic search.
- Critical appraisal. Finally all full papers relevant to each clinical question were appraised using the methodology described in the NICE Guideline Development Methods manual.

It should be noted that most of the published research on cancer topics focuses on clinical evaluations of treatment; little direct research has been carried out on the organisation and delivery of services.

All the evidence reviews used to inform the manual are summarised in the document 'Improving outcomes for people with sarcoma: the research evidence', which includes details of all the studies appraised. This document is available on CD ROM, a copy of which is included on the inside cover of the manual.

Additional complementary research, designed to quantify the potential cost of major changes in services, was carried out by the Centre for Economics and Policy in Health, Institute of Medical and Social Care Research (IMSCAR), at the University of Bangor. This work involves literature searching, interviews with clinicians and managers, and analyses of costs.

The writing of the guidance manual was coordinated by the Chair and Clinical Lead of the GDG in accordance with all members of the GDG, assisted by staff at the NCC-C.

The production of this guidance was funded by NICE, and has been subject to the full NICE consultation process.

People and organisations involved in production of this guidance

4.1 Members of the Guidance Development Group

4.2 Organisations invited to comment on guidance development

4.3 People carrying out literature reviews and complementary work

4.4 Expert advisers to the Guidance Development Group

4.5 Members of the Guideline Review Panel

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Appendix 4.1

Members of the Guidance Development Group (GDG)

GDG Chair

Dr Joe Kearney Director of Public Health, Dacorum,
Watford and Three Rivers PCTs

GDG Lead Clinician

Mr Robert Grimer Consultant Orthopaedic Surgeon, Royal
Orthopaedic Hospital, Birmingham

Group Members

Dr Albert Benghiat Cancer Network Director and Clinical
Oncologist, Leicester Royal Infirmary

Ms Janine Broadbent Patient/Carer Representative, Sarcoma UK

Dr Wyn Davies General Practitioner Principal, Cardiff

Mrs Merian Denning Senior I Physiotherapist, Christie Hospital
NHS Trust, Manchester

Mrs Joy Dowd Macmillan Clinical Nurse Specialist,
Freeman Hospital, Newcastle upon Tyne

Dr Janet Glencross Director of Public Health, Lincolnshire
South West Primary Care Trust

Dr Jeremy Jenkins Consultant Radiologist, Central Manchester
and Manchester Children's University
Hospital NHS Trust and Honorary Clinical
Senior Lecturer, University of Manchester

Professor Ian Judson	Professor of Cancer Pharmacology/Honorary Consultant Medical Oncologist, The Institute of Cancer Research and Royal Marsden NHS Trust
Dr Michael Leahy	Consultant Medical Oncologist, Christie Hospital NHS Trust, Manchester
Professor Archie Malcolm	Consultant Pathologist, Royal Shrewsbury Hospital and Professor of Pathology, Keele University
Professor Malcolm Reed	Professor of Surgical Oncology, University of Sheffield and Honorary Consultant Surgeon, Royal Hallamshire Hospital, Sheffield
Dr Julia Riley	Consultant in Palliative Medicine, The Royal Marsden Hospital, London
Mrs Judith Robinson	Patient/Carer Representative, Sarcoma UK
Dr Martin Robinson	Senior Lecturer, Honorary Consultant in Clinical Oncology, Weston Park Hospital, Sheffield
Mr J Meirion Thomas	Consultant Surgeon, The Royal Marsden Hospital, London
Roger Wilson	Patient/Carer Representative, Sarcoma UK

Appendix 4.2

Organisations invited to comment on guidance development

Addenbrooke's NHS Trust

Anglesey Local Health Board

Association for Palliative Medicine of Great Britain and Ireland

Association of Hospice and Specialist Palliative Care Social Workers

Association of Surgeons of Great Britain and Ireland

Association of the British Pharmaceuticals Industry (ABPI)

Association of Upper Gastrointestinal Surgeons of Great Britain and Ireland (AUGIS)

Bard Limited

Bath and North East Somerset PCT

Baxter Oncology

Bedfordshire & Hertfordshire NHS Strategic Health Authority

Boehringer Ingelheim Ltd

Brighton & Sussex University Hospitals Trust

British Association for Counselling and Psychotherapy

British Association for Dermatological Surgery

British Association of Art Therapists

British Association of Head and Neck Oncologists

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British Association of Oral and Maxillofacial Surgeons
British Association of Otolaryngologists, Head & Neck Surgeons
British Association of Plastic Surgeons
British Bone and Soft Tissue Tumour Panel
British National Formulary (BNF)
British Oncology Pharmacy Association
British Orthopaedic Association
British Psychological Society
British Psychosocial Oncology Society
British Society for Dermatopathology
British Society of Paediatric Radiology
British Society of Skeletal Radiology
BUPA
Cancer and Leukaemia in Childhood (UK)
Cancer Research UK
Cancer Services Collaborative 'Improvement Partnership' (CSCIP)
Cancer Services Co-ordinating Group
Cancer Voices
CancerBACUP
Chartered Society of Physiotherapy
Children's and Adolescent Cancer Partnership (CACP)
Clatterbridge Centre for Oncology NHS Trust
College of Occupational Therapists
Coloplast Limited
Countess of Chester Hospitals NHS Trust

Department of Health
Eisai Limited
Faculty of Public Health
Guerbet Laboratories Ltd
Healthcare Commission
Help Adolescents with Cancer
Hull and East Yorkshire NHS Trust
Intra-Tech Healthcare Ltd
Joint Committee on Palliative Medicine
Leeds Teaching Hospitals NHS Trust
Limbless Association
Macmillan Cancer Relief
Marie Curie Cancer Care
Medical Research Council Clinical Trials Unit
Medicines and Healthcare Products Regulatory Agency (MHRA)
Middlesbrough Primary Care Trust
National Alliance of Childhood Cancer Parent Organisations
National Cancer Alliance
National Cancer Network Clinical Directors Group
National Cancer Research Institute – Sarcoma Clinical Studies Group
National Cancer Research Institute (NCRI) Clinical Studies Group
National Council for Disabled People, Black, Minority and Ethnic
Community (Equalities)
National Patient Safety Agency
National Public Health Service – Wales

Neurofibromatosis Association

NHS Direct

NHS Health and Social Care Information Centre

NHS Modernisation Agency

NHS Quality Improvement Scotland

North of England Bone and Soft Tissue Tumour Service

Northumberland Care Trust

Novartis Pharmaceuticals UK Ltd

Nuffield Orthopaedic Centre NHS Trust

Pfizer Limited

PharmaMar

Princess Alexandra Hospital NHS Trust

Richmond & Twickenham PCT

Robert Jones & Agnes Hunt Orthopaedic & District Hospital
NHS Trust

Royal College of Anaesthetists

Royal College of General Practitioners

Royal College of General Practitioners Wales

Royal College of Nursing (RCN)

Royal College of Paediatrics and Child Health

Royal College of Pathologists

Royal College of Physicians of London

Royal College of Psychiatrists

Royal College of Radiologists

Royal College of Surgeons of England

Royal College Patient Liason Groups

Royal Liverpool Children's NHS Trust

Royal Marsden Hospital NHS Trust

Royal National Orthopaedic Hospital NHS Trust

Royal Pharmaceutical Society of Great Britain

Royal Society of Medicine

Royal West Sussex Trust

Sarcoma UK

Scottish Bone and Soft Tissue Sarcoma Network

Scottish Intercollegiate Guidelines Network (SIGN)

Sheffield South West Primary Care Trust

Sheffield Teaching Hospitals NHS Trust

Society and College of Radiographers

South Warwickshire General Hospitals NHS Trust

South West Cancer Intelligence Service

South West London Strategic Health Authority

Tameside and Glossop Acute Services NHS Trust

Teenage Cancer Trust

Thames Valley Strategic Health Authority

UK Children's Cancer Study Group

University College London Hospital NHS Trust

University Hospital Birmingham NHS Trust

Welsh Assembly Government (formerly National Assembly for Wales)

Wessex Cancer Trust

West Lincolnshire PCT

People carrying out literature reviews and complementary work

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Expert advisers to the Guidance Development Group

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Dr Jai Kulkarni	Consultant and Honorary Lecturer in Rehabilitation Medicine, University Hospitals of South Manchester
Mr Satvinder Mudan	Consultant Surgeon, The Royal Marsden Hospital

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Members of the Guideline Review Panel

Chair

John Hyslop

Members

Graham Archard

Tony Donovan

Mark Emberton

Patricia Fairbrother

Stephen Karp

Glossary of terms

Adjuvant chemotherapy

Chemotherapy treatment that is given in addition to the main cancer treatment.

Adjuvant therapy

Additional treatment that is added to increase the effectiveness of the main treatment.

Allied health professional (AHP)

One of the following groups of healthcare workers: physiotherapists, occupational therapists, art therapists, chiropodists/podiatrists, dietitians, drama therapists, music therapists, orthoptists, paramedics, prosthetists/orthotists, diagnostic radiographers, therapeutic radiographers, speech and language therapists.

Asymptomatic

Without obvious signs or symptoms of disease. In early stages, cancer may develop and grow without producing symptoms.

Axial skeleton

The jointed bones of the head and vertebral column.

Benign

Not cancerous; not malignant.

Biopsy

Removal of a sample of tissue or cells from the body to assist in the diagnosis of a disease.

Bone sarcoma

Sarcomas, such as osteosarcoma, affecting the bone.

Cancer networks

The organisations for cancer services to implement the NHS Cancer Plan, bringing together health service commissioners and providers, the voluntary sector and local authorities. There are currently 34 cancer networks in England and three in Wales, covering between 600,000 and 3 million population (two-thirds serve a population of between 1 and 2 million people).

Carcinoma

Cancer of the lining tissue that covers all the body organs. Most cancers are carcinomas.

Chemotherapy

The use of drugs that kill cancer cells, or prevent or slow their growth.

Chondrosarcoma

A malignant tumour derived from cartilage tissue.

Clinical oncologist

A doctor who specialises in the treatment of cancer patients, particularly through the use of radiotherapy, but may also use chemotherapy.

Cohort study

Research studies in which groups of patients with a particular condition or specific characteristic are compared with matched groups who do not have it.

Computed tomography (CT)

An X-ray technique that produces cross-sectional images.

Connective tissue

Forms the supportive and connective structures of the body.

Counselling

Counselling takes place when a counsellor sees a client in a confidential setting to explore a difficulty the client is having, distress they may be experiencing or their dissatisfaction with life.

Curative

Aiming to cure a disease.

Cytogenetics

The study of chromosomes and chromosomal abnormalities.

Diagnostic radiographer

A healthcare professional trained in the technique of obtaining images of various parts of the body, working closely with a radiologist.

Diaphysis

The shaft of a long bone.

Dietitian

The healthcare professional responsible for the planning and managing of the patient's diet in hospital and providing dietary advice for a wide range of medical conditions.

Enchondromas

A cartilaginous tumour growing from the interior of a bone.

Endoprostheses

Artificial devices placed within the body to replace a natural function (usually referring to bone replacements such as for the hip, knee, etc.).

Epidemiology

The study of populations in order to determine the frequency and distribution of disease and to measure risks.

Ewing's sarcoma

A type of bone cancer that usually forms in the middle (shaft) of large bones. It occurs most frequently in children and young adults.

Excise/excision

Removal by surgery.

Fascia

Flat layers of fibrous tissue that separate different layers of tissue.

Fibrosarcoma

A type of soft tissue sarcoma that begins in fibrous tissue, which holds bones, muscles, and other organs in place.

Fractionation

Dividing the total dose of radiation therapy into several smaller, equal doses delivered over a period of several days.

Gastrointestinal stromal tumour (GIST)

An unusual and specific type of tumour that usually begins in cells in the wall of the gastrointestinal tract (stomach, small bowel).

Gastrointestinal tract

The part of the digestive system that includes the mouth, oesophagus, stomach, and intestines.

Gynaecology

A branch of medicine dealing with the diagnosis and treatment of disorders affecting the female reproductive organs.

Haematemesis

The vomiting of blood.

Hereditary multiple exostoses

A genetic condition in which bones develop multiple abnormal lumps.

High-grade

These cancers tend to grow more aggressively, are more malignant, and have the least resemblance to normal cells.

Histological

Relating to the study of cells and tissue on the microscopic level.

Histopathologist

A doctor who specialises in examining tissue samples microscopically in order to make a diagnosis and ensure tumour excision is complete.

Histopathology

The study of microscopic changes in diseased tissues.

Holistic

Looking at the whole system rather than just concentrating on individual components.

Imatinib

A drug used in the treatment of patients with metastatic or inoperable GIST.

Immunohistochemistry

A technique that uses antibodies to show up specific proteins in tissues seen down a microscope.

Isolated limb perfusion

Chemotherapy treatment in which blood is taken from a patient, pumped through a machine that adds anticancer drugs to the blood, then returned to the limb being treated.

Key worker

Person who, with the patient's consent and agreement, takes a key role in coordinating the patient's care and promoting continuity, ensuring the patient knows who to access for information and advice.

Laparotomy

General term for abdominal surgery.

Late effect

A side effect of radiotherapy or chemotherapy that occurs some months or years after treatment.

Leiomyosarcoma

A malignant tumour of smooth muscle origin.

Li-Fraumeni syndrome

An inherited family trait carrying an increased risk of cancer during childhood and early adulthood.

Lymph node dissection

A surgical procedure in which lymph nodes are removed and examined to see whether they contain cancer. Also called lymphadenectomy.

Lymphadenectomy

See lymph node dissection.

Lymphoedema

A condition in which excess fluid collects in tissue and causes swelling. It may occur in the arm or leg after lymph vessels or lymph nodes in the underarm or groin are removed or treated with radiation.

Magnetic resonance imaging (MRI)

A non-invasive method of imaging, which allows the form and metabolism of tissues and organs to be visualised (also known as nuclear magnetic resonance).

Malignant

Cancerous. Malignant tumours can invade and destroy nearby tissue and spread to other parts of the body.

Malignant fibrous histiocyoma

A soft tissue sarcoma that usually occurs in the limbs, most commonly the legs, and may also occur in the abdomen.

Medical oncologist

A doctor who treats cancer patients through the use of chemotherapy, and for some tumours, immunotherapy.

Melaena

Abnormally dark tarry faeces containing blood.

Mesenchymal

Relating to embryonic tissue of mesodermal origin.

Metastases

Cancerous tumours in any part of the body that have spread from the original (primary) origin.

Metastatic disease

The spread of a disease from the organ or tissue of origin to another part of the body.

Molecular pathology

New techniques for identifying molecular abnormalities in the DNA of tumour cells.

Morbidity

The state of being diseased.

Mortality

Either (1) the condition of being subject to death; or (2) the death rate, which reflects the number of deaths per unit of population in any specific region, age group, disease or other classification, usually expressed as deaths per 1000, 10,000 or 100,000.

Mutation analysis

Testing for the presence of a specific mutation, a specific type of mutation or set of mutations.

Neoplasm

An abnormal mass of tissue that results from excessive cell division – a tumour.

Neurofibromatosis

A genetic condition in which people develop multiple, benign tumours of nerve tissue.

Neutropenic sepsis

Life-threatening infection made more severe by the patient's having a very low level of white blood cells.

Observational study

A non-randomised study that observes the characteristics and outcomes over time of subjects who do and do not take a particular therapy.

Occupational therapist

A healthcare professional trained to help people who are ill or disabled learn to manage their daily activities.

Ollier's disease

The benign growth of cartilage in the metaphyses of several bones.

Oncology

The study of the biological, physical and chemical features of cancers. Also the study of the causes and treatment of cancers.

Oncologist

A doctor who is trained to treat patients with chemotherapy (medical oncologist), radiotherapy or both (clinical oncologist).

Orthopaedic surgeon

A doctor who specialises in the surgery of bones.

Orthosis

A device that is used to protect, support or improve function of parts of the body that move.

Orthotist

A skilled professional who fabricates orthotic devices that are prescribed by a physician.

Osteochondroma

A non-cancerous tumour made up of bone and cartilage.

Osteosarcoma

A cancer of the bone that usually affects the large bones of the arm or leg. It occurs most commonly in young people and affects more males than females.

Paediatric oncologist

An oncologist who specialises in the treatment of children.

Paget's disease

A disease of the bone.

Palliative

Anything that serves to alleviate symptoms due to the underlying cancer but is not expected to cure it.

Palpable mass

A mass that can be felt by the doctor.

Pathologist

A doctor who examines cells and identifies them. The pathologist can tell where a cell comes from in the body and whether it is normal or a cancer cell. If it is a cancer cell, the pathologist can often tell what type of body cell the cancer developed from. In a hospital practically all the diagnostic tests performed with material removed from the body are evaluated or performed by a pathologist.

Pathology

A branch of medicine concerned with disease, especially its structure and its functional effects on the body.

Physiotherapist

A specialist trained in using exercise and physical activities to condition muscles and improve the level of activity.

Positron emission tomography (PET)

A highly specialised imaging technique using a radioactive tracer to produce a computerised image of body tissues to find any abnormalities. PET scans are sometimes used to help diagnose cancer and investigate a tumour's response to treatment.

Prognosis

A prediction of the likely outcome or course of a disease; the chance of recovery or recurrence.

Prosthetist

A specialist who makes and fits artificial limbs and similar devices.

Prosthetics

An artificial device used to replace a missing part of the body.

Psychological support

Professional support that can help people with a wide range of psychological problems such as anxiety and depression, and can provide emotional assistance during times of distress.

Psychologist

A specialist who can talk with patients and their families about emotional and personal matters, and can help them make decisions.

Psychosocial

Concerned with psychological influence on social behaviour.

Quaternary

Fourth level. Relating to medical treatment provided at a very specialist institution.

Radiologist

A doctor who specialises in creating and interpreting pictures of areas inside the body using X-rays and other specialised imaging techniques. An interventional radiologist specialises in the use of imaging techniques for treatment, for example catheter insertion for abscess drainage.

Radiology

The use of radiation (such as X-rays, ultrasound and magnetic resonance) to create images of the body for diagnosis.

Radiotherapy (radiation treatment)

The use of ionising radiation, usually X-rays or gamma rays, to kill cancer cells and treat tumours.

Randomised controlled trial (RCT)

A type of experiment that is used to compare the effectiveness of different treatments. The crucial feature of this form of trial is that patients are assigned at random to groups which receive the interventions being assessed or control treatments. RCTs offer the most reliable (i.e. least biased) form of evidence of effectiveness.

Reconstructive surgery

Surgery that is done to reshape or rebuild (reconstruct) a part of the body changed by previous surgery.

Resection

Removing tissue from the body by surgery.

Retinoblastoma

An eye cancer that most often occurs in infants and young children.

Retroperitoneum

The space behind the peritoneum (a membrane that lines the entire abdominal wall of the body).

Retroperitoneal sarcoma

A sarcoma that develops in the tissues at the back of the abdominal cavity.

Rhabdomyosarcoma

A malignant tumour of muscle tissue.

Sarcoma

A cancer of the bone, cartilage, fat, muscle, blood vessels, or other connective or supportive tissue.

Soft tissue

Refers to muscle, fat, fibrous tissue, blood vessels, or other supporting tissue of the body.

Soft tissue sarcoma (STS)

A cancer of the soft tissues of the body.

Spindle cell sarcoma

A type of connective tissue cancer in which the cells are spindle-shaped when examined under a microscope.

Subcutaneous tumour

A tumour beneath the skin.

Synovial sarcoma

A malignant tumour that begins as a soft swelling and often metastasises.



Systematic review

A review of the literature carried out in order to address a defined question and using quantitative methods to summarise the results.

Tertiary

Third level. Relating to medical treatment provided at a specialist institution.

Therapeutic radiographer

The role of the therapeutic radiographer is to work closely with other specialists, to deliver the radiotherapy as prescribed, to give patients information and support, and to discuss possible side effects and care.

Thoracic

Relating to the chest.

Toxicity

Refers to the undesirable and harmful side effects of a drug.

Truncal

Relating to the trunk of the body or to any arterial or nerve trunk.

Ultrasound

A non-invasive technique using ultrasound waves (high-frequency vibrations beyond the range of audible sound) to form an image.

Unresectable

A tumour or mass that cannot be removed by surgery.

Viscera

The internal organs enclosed in a body cavity such as the abdomen, chest or pelvis.

X-ray

A photographic or digital image of the internal organs or bones produced by the use of ionising radiation.

Abbreviations

AHP	allied health professional
BSRM	British Society of Rehabilitation Medicine
CNS	clinical nurse specialist
CPA	clinical pathology accreditation
CPD	continuing professional development
CT	computed tomography
DGH	district general hospital
DSC	disablement service centre
EQA	external quality assurance
FTE	full-time equivalent
GDG	Guidance Development Group
GI	gastrointestinal
GIST	gastrointestinal stromal tumour
GP	general practitioner
HES	hospital episode statistics
HME	hereditary multiple exostoses
ICCC	International Classification of Childhood Cancer
ICD	International Classification of Diseases
LHB	local health board
MDT	multidisciplinary team
MRI	magnetic resonance imaging
NCC-C	National Collaborating Centre for Cancer
NCRI	National Cancer Research Institute
NCRN	National Cancer Research Network
NICE	National Institute for Health and Clinical Excellence
NSCAG	National Specialist Commissioning Advisory Group
NSF	National Service Framework

ONS	Office for National Statistics
PARC	Prosthetic and Amputee Rehabilitation Centre
PCT	primary care trust
PDP	personal development plan
PEDW	patient episode database, Wales
PET	positron emission tomography
SSP	specialist sarcoma pathologist
STS	soft tissue sarcoma
UKCCSG	United Kingdom Children's Cancer Study Group
US	ultrasound
WHO	World Health Organization

