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# Guidance on Cancer Services

## Improving Outcomes for People with Sarcoma

### The Manual

The paragraphs are numbered for the purposes of consultation. The final version will not contain numbered paragraphs.

## NICE Stakeholder Consultation Version

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## Foreword

1. This 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.
2. 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 two years, especially the patient representative members whose sensible reflections of 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**

## Key Recommendations

3. All patients with a confirmed diagnosis of bone or soft tissue sarcoma should have their care supervised by or in conjunction with a sarcoma MDT.
4. Commissioners should ensure that cancer networks arrange diagnostic services for the investigation of patients with potential sarcomas (as defined by the urgent referral criteria) at designated diagnostic clinics.
5. All patients with a provisional histological or radiological diagnosis of bone or soft tissue sarcoma should have their diagnosis reviewed by a specialist sarcoma pathologist or radiologist. A formal system for second opinions and review of difficult cases should be funded.
6. A soft tissue sarcoma MDT should meet minimum criteria (as defined in Chapter 5) and manage at least 100 new soft tissue sarcoma patients per year. If a sarcoma MDT manages both bone and soft tissue it needs to manage at least 50 new bone sarcoma patients per year and at least 100 new soft tissue sarcomas patients per year. If a sarcoma MDT only manages bone sarcomas, then it should manage at least 100 new bone sarcoma patients per year.
7. A key worker, who will be a member of the sarcoma MDT should be allocated to each sarcoma patient.
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 site specific skills in consultation with the sarcoma MDT.

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9. 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.
10. All sarcoma MDTs should participate in national audit, data collection and training and be encouraged to enter patients into clinical trials.
11. Patients with functional disabilities should have access to appropriate support and rehabilitation services.
12. The National Specialist Commissioning Advisory Group should consider funding a number of designated centres for management of abdominal and pelvic soft tissue sarcomas.

## Introduction

13. This guidance advises commissioners on how to improve the care of patients with bone and 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 involve 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 within and across cancer networks to achieve the important improvements in care these patients require.
  
14. 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. In addition the Guidance Development Group (GDG) consists of a wide range of experienced health professionals and patients, the value of whose advice should not be underestimated. We believe that if this guidance is implemented, important and worthwhile changes will occur nationally in the management of these patients.
  
15. 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

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- soft tissue sarcomas and the large number of hospitals currently involved in delivering care to these patients.
16. The patient perspective follows. We have attempted to be 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.
  17. 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 draft NICE guidelines on *Referral for Suspected Cancer* were our starting point. 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. We have described the diagnostic pathways that patients with bone or soft tissue sarcomas should follow and hope that these recommendations will make a significant difference.
  18. Expertise in radiology and histopathology are 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 so 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.
  19. 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

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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 we believe that 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 co-operatively to address this requirement.

20. We have tried to ensure 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 health professionals within cancer networks address the recommendations.
21. 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 think we have managed this balance by recommending the most appropriate patient groups to the most appropriate place of treatment.
22. This diverse group of tumours can occur almost anywhere in the body. We think we have addressed all the main areas where MDTs from different disciplines will need to work closely together. We have attempted to support 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. For patients with sarcomas at some sites it should be recognised that there is a need for even more specialisation than is



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recommended here. We have not given definitive advice other than to highlight this issue and trust that, as managed sarcoma care develops in England and Wales, such sub-specialisation will be established. We have paid particular attention to gastro-intestinal 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.

23. The 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, then both the support to the individual patient with a key worker and 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*. We have tried to avoid too much duplication.

24. 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

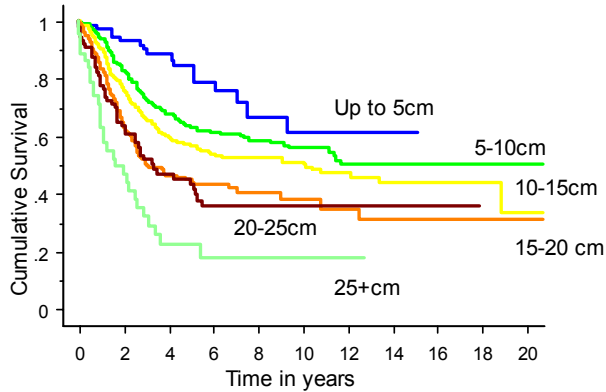
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commissioners are unlikely to provide the necessary increases in resource this patient group requires.

## Chapter 1 - Background

25. Sarcomas are a rare and diverse group of cancers thought to have a common embryological origin. They arise from cells that comprise 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 23<sup>rd</sup> most common cancer type and bone tumours the 27<sup>th</sup> most common type. If combined sarcomas would still only be the 21<sup>st</sup> most common cancer type [1].
26. During a working lifetime a 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. 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.
27. Delays in diagnosis for both bone and STS are common. The median size on presentation for both bone and STS is 10 cm – earlier diagnosis would undoubtedly lead to improved outcomes both in terms of survival and 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.**



### ***Incidence of Sarcomas in England and Wales***

28. The coding system used for cancers is the International Classification of Diseases, currently in its 10th edition (ICD-10). The ICD-10 classifies cancers to specific body sites with codes for tumours of bone and for tumours of connective and soft tissue.

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

29. Ideally all cancers, of whatever type, are recorded by the Cancer Registries in England and Wales and over 90% ascertainment is currently reported [2]. For the five year period between 1996 and 2000 there were 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 E&W 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

30. 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 in the number of soft tissue sarcomas 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 E&W 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 ONS and Welsh Cancer Intelligence and Surveillance Unit

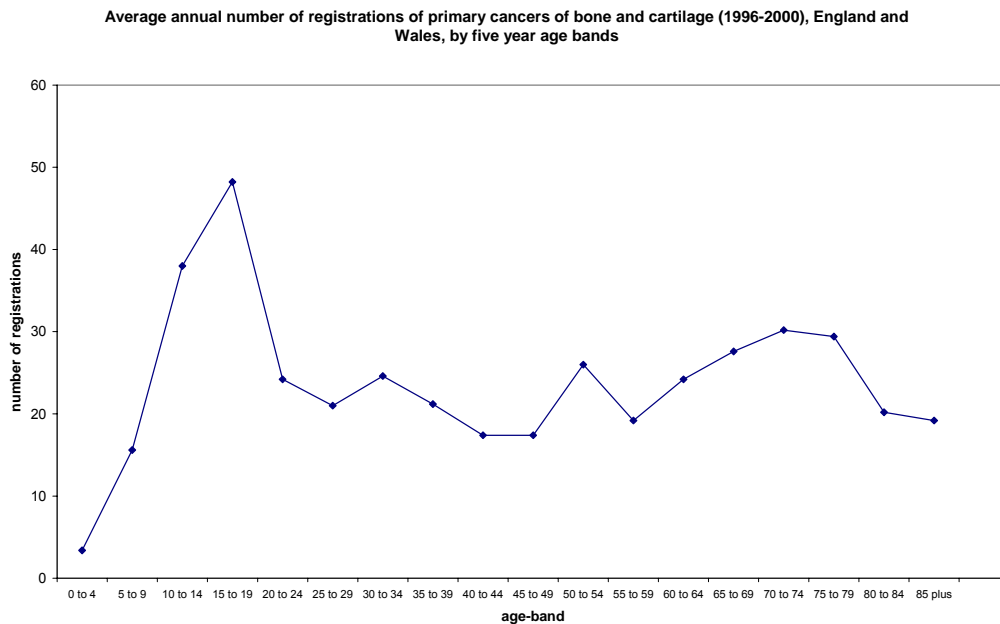
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31. Five year data from the East Anglia Cancer Registry was reviewed for this guidance and it was found that only 53% of soft tissue sarcomas were coded to ICD10 C49. Other sites included the uterus, gastro-intestinal tract and the retroperitoneum.
32. If coding practices are similar throughout England and Wales then approximately 2000 soft tissue sarcomas, in all sites, might be expected each year.
33. It is important to note here that the historical cancer registry data do not take into account the recent advances in the classification of gastro-intestinal 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 [3].

### ***Bone Sarcomas***

34. Bone sarcomas are estimated to account for 0.2% of all malignant tumours, but represent 4% of all malignancy in children aged up to 14 years. 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.
35. 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 sixty. They are more common in males than in females. Figure 2 shows the age specific incidence for primary bone cancers.

Figure 2



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

- *Osteosarcomas*

37. The most common primary malignant bone tumour. It occurs predominantly in patients younger than twenty, in whom 80% of tumours occur in long bones of the extremities. In the older age group osteosarcomas may arise secondary to radiation or Paget's disease.

- *Chondrosarcomas*

38. The incidence of this type of malignant bone tumour increases gradually with age. More than 50% of these tumours occur in the long bones of the extremities. They may also occur in the pelvis and ribs.

- *Ewing's sarcoma*

39. The major peak for age-specific incidence occurs in the second decade of life with a rapid decrease after the age of 20 years. These tumours are reported to occur almost exclusively in the white population. They typically

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arise in the axial skeleton (pelvis, scapula, rib) or in the diaphysis (main or mid section) of long bones.

- *Spindle cell sarcomas*

40. There are a variety of other rare sarcomas of bone e.g. fibrosarcoma, malignant fibrous histiocytoma and leiomyosarcoma, which behave just like osteosarcoma but typically arise in an older population.

### **Risk factors for bone sarcomas**

41. 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***

42. 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).

#### ***Pre-cursor conditions***

43. There are several pre-cursor 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% to 10% of patients with severe Paget's disease.

44. 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 Mafucci's disease, are also at an increased risk.



***Radiation***

45. 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).

**Treatment of bone sarcomas**

46. 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 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 physiotherapy and rehabilitation to recover optimal personal and social functioning, including return to work. Some patients require lifelong provision of orthotic and/or prosthetic appliances.

47. Chemotherapy regimens are used for bone sarcoma and are among the most complex in adult oncology practice. Adjuvant chemotherapy (given both pre-operatively and post-operatively) contributes significantly to long term survival for patients with Ewing's sarcoma and osteosarcomas.

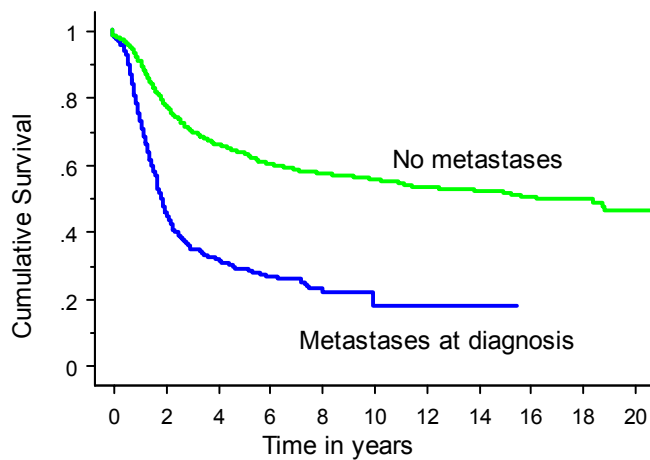
48. Radiotherapy is a key part of curative treatment for some patients with Ewing's sarcoma and is a valuable part of palliative therapy for other patients with bone sarcoma. Radiotherapy is typically delivered by fractionation of the total dose over four to six weeks with daily attendances for treatment.

**Prognosis and survival of bone sarcomas**

49. 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 five year relative

survival rates increased from 29% in 1971-75 to 51% in 1986-90 [4]. 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.**



50. The European cancer registry based study on survival and care of cancer patients [5] aims to describe survival differences between European populations and the reasons for them. For adults diagnosed between 1990-1994 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 quality of data available in different countries.)

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

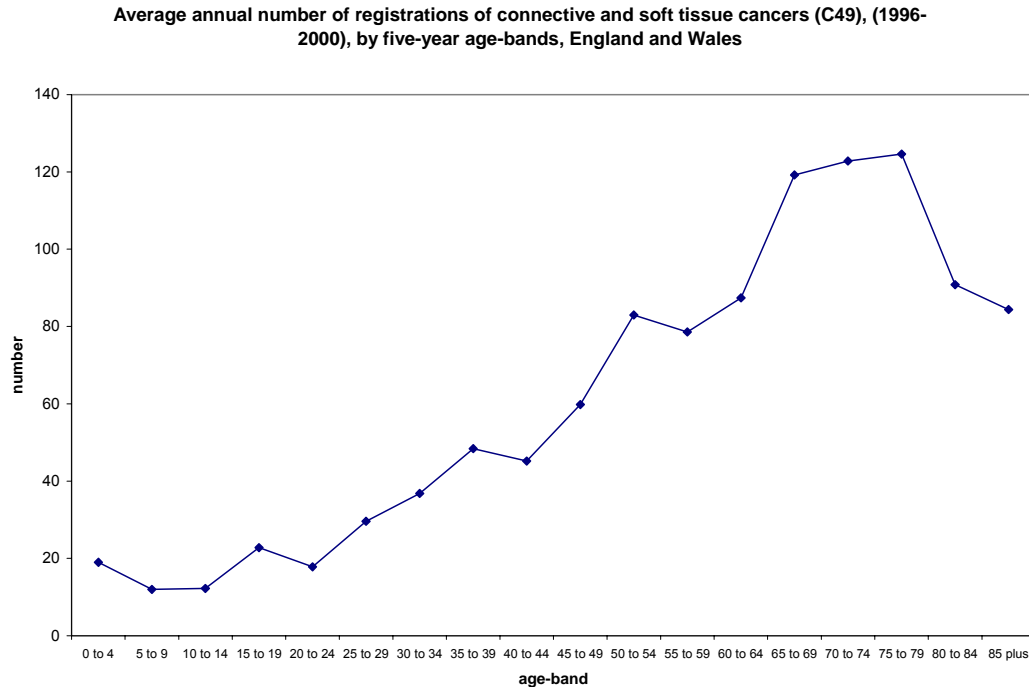
Country	MEN Age-standardised relative survival (%) and 95% CI	WOMEN Age-standardised relative survival (%) and 95% CI
England	51.4 (46.9-56.3)	54.8 (49.8-60.3)
Europe	53.0 (48.0-58.4)	56.3 (51.5-61.6)

Data from EUROCARE-3

### ***Soft Tissue Sarcomas (STS)***

51. Soft tissue sarcomas (STS) account for about 1% of all malignant tumours. 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



- *Extremity and superficial trunk*

52. The majority of patients with STS of the extremities and superficial trunk present with a painless mass. It can be difficult to differentiate a benign from a malignant mass but draft urgent referral guidelines [6] have been produced by NICE to identify patients more likely to have a malignant tumour.

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* (draft for consultation)

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- *Retroperitoneum*

53. Most patients present with an abdominal mass, with half reporting pain at presentation. Due to the space available in the retroperitoneum these tumours may often grow to a substantial size before presenting and their overall prognosis is worse than extremity sarcomas.

- *Viscera*

54. Sarcomas of the viscera present with signs and symptoms particular to the organ of origin. For example, gastro-intestinal stromal tumours (GIST), which occur primarily in the middle aged and older population, present with upper abdominal pain in 40-50% of cases. 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.

- *Head and Neck*

55. 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. They can present as a lump, with problems relating to compression of surrounding anatomy such as the orbit or pharynx. Surgery and radiotherapy is difficult due to the proximity of important anatomy in this area.

### **Risk factors of STS**

56. 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.

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

58. Prior radiotherapy can also cause late development of soft tissue sarcomas.

### **Treatment of STS**

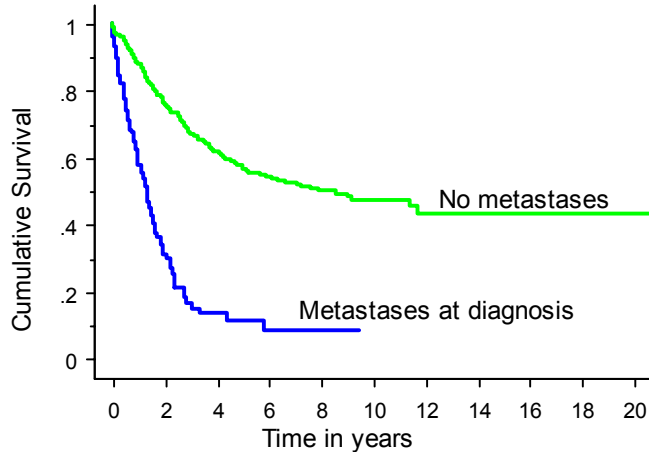
59. The treatment of STS is largely surgical – excising the tumour with as wide a margin of surrounding normal tissue as possible. For patients with large and high grade tumours, radiotherapy will also usually be used. Chemotherapy is principally used for treating specific STS (soft tissue Ewing's tumour, rhabdomyosarcomas and in children with STS) 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 [7].

### **Prognosis and survival of STS**

60. Five year survival is between 50% and 60% for soft tissue sarcomas as a group, however there is wide variation depending on anatomical site and histological features of the tumours. Data from EURO CARE-3 [8] shows 5 year survival for tumours coded to 'soft tissue' in England is not significantly different from Europe as a whole.

61. 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. Patients with retroperitoneal sarcomas have a poorer prognosis largely because these tumours present so late.

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



## ***Current Services for Sarcomas***

### **Bone sarcomas**

62. The diagnosis and surgical treatment of primary bone tumours are very complex and the two supraregional bone tumour treatment centre, 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.

63. 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 endoprosthetic replacement. In 2005 NSCAG therefore widened the definition and will commission services for “the diagnosis and

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treatment of primary bone tumours". Other units including Newcastle, Oxford, Bristol, and Oswestry are now contracted to supply this service.

64. 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) (1997/98-2001/02) identified 33 trusts that had more than 50 episodes coded to medical and/or clinical oncology for the treatment of primary bone tumours.
65. Most primary malignant bone tumours occur in adolescents or children and these patients will receive their non-surgical treatment at a children's or young adult oncology centre. They will however require complex surgery, carried out at an NSCAG centre.

### **Soft tissue sarcomas**

66. 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.
67. A review of 5 years of 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 eighteen trusts reported that they performed planned



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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 gynaecological sarcomas and those of the gastro-intestinal tract are often managed by those specialty multidisciplinary teams.

68. 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**

69. 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. The precise definition of what constitutes a sarcoma treatment centre, both in terms of standards, number of patients treated and of staffing has not been consistently defined.

70. 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**

71. There is no established system of training 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. Some centres do not have anyone with recognised training involved in the treatment of sarcomas.

## Patient support

72. 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 which must be addressed. Sarcoma surgery is frequently disabling or disfiguring and although fewer patients face amputation than in the past they require life-time 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 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 soft tissue sarcomas.
- Significant numbers of patients especially with STS, are probably not managed by a multidisciplinary team.
- Most bone tumours occur in children and young people.

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## Chapter 2 - Patient perspectives

73. 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. There therefore needs to be a clear focus on access to relevant, high-quality and timely information and emotional support from family and health professionals.
74. 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.
75. 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.
76. 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 patients wish to receive the best possible treatment that's available to them and will cope with the travel issues as a secondary pressure if necessary.

## ***Information***

77. 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 their specific situation
- leaflets offering generic treatment information (e.g. about chemotherapy)
- audio, video or CD resources
- the Internet
- telephone helpline with access to a specialist nurse or other healthcare professional.

78. 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 audio tape, and may need to be translated into languages other than English, or made available in large print versions.

79. 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 which the patient can ask their own doctor about.

80. The Internet presents a range of problems with regard to information quality. A patient using the Internet will inevitably come across sites which 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***

81. 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.

82. 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 groups where decisions may be traditionally taken within the family on behalf of a patient.

### ***Choice and Decision Making***

83. 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 which 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.

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84. The prospect of participating in a clinical trial is an additional burden for patients. As many sarcoma trials are for drug treatments in a palliative context it is important that the choices available are made clear to patients who are eligible for a trial.

### ***GP Information***

85. General practitioners 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 sarcoma patient 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 could also be referred to in communication between treatment centres and the patients’ GPs.

## **A. Recommendations**

### ***Diagnosis***

86. Communicating a diagnosis or other significant news should be undertaken by a senior doctor or specialist nurse who has enhanced skills (as defined in the NICE guidance on Improving Supportive and Palliative Care for Adults with Cancer). This should be undertaken face-to-face unless there is specific agreement with the patient about receiving confirmation of a preliminary diagnosis by telephone or in writing.

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

88. Patients should be offered a permanent written record of their diagnosis and of any important points relating to the consultation. Their key worker

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should be identified in writing together with their contact points and this information should also be supplied to their GP.

### ***Information***

89. 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 patients' diagnosis or disease stage.

90. Information should be provided in printed format and supported by information about access to on-line 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 they can understand.

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

92. Table 4 maps the scope of the information which should be made available to patients at each stage in the disease/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 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 post if the diagnosis is given by telephone
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	Sarcoma treatment centre (see Chapter 5) by post
	Specific information on the diagnosis and the proposed treatment (if known and if not given by diagnostic clinic)	Local arrangements can apply
On any treatment decision	Generic information on that treatment (surgery, radiotherapy, chemotherapy) and any tests or imaging procedures which 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	Information on the new sarcoma treatment centre. Identification of key worker	New sarcoma treatment centre by post

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After surgery or other treatment	Specific information on individual follow-up procedure, self monitoring information, healthcare support and sarcoma specific support	Sarcoma treatment centre by post or face-to-face as appropriate
	Confirmation of the named key worker for that patient together with contact details	
	Specific information on support for prosthetic limbs or endoprosthesis implants	
	Details of generic local and national support groups and other support resources	Sarcoma treatment centre or patient support centre, face-to-face or by post
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 proposed trial.	Sarcoma treatment centre face-to-face. Specific 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 and GP

93. Generic information may include publications from Cancer BACUP or other voluntary sector providers. Where such generic information is relied upon and is not immediately available at the diagnostic clinic/sarcoma

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treatment centre the patient should be given the telephone number of the agency and the specific title of the relevant booklet.

94. Clinical trials which are not being conducted at the patient's own treatment centre should be offered to the patient at another treatment centre of their choice if possible.

95. Details of clinical trials for sarcoma should be available at every sarcoma treatment centre.

### ***Support***

96. Patients should be offered appropriate support as shown below:

- Psychological support.
- Spiritual support.
- Social support through contact with others facing similar situations – self help groups.
- Practical healthcare support relating to treatment.
- Benefits advice.

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

### ***General***

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

99. In the event of a significant delay or alteration in diagnosis which affects a patient's management then a 'significant event analysis' should be undertaken and the lessons learnt from this fed back both to relevant clinicians and the patient informed.

## **B. Anticipated benefits**

100. Provision of clear, well communicated and timely information will improve the patients' 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.
101. Patients will benefit by both helping and being helped by others with a similar condition.
102. There will be a clear identification of what information should be provided, by whom and when.

## **C. Evidence**

### ***Communication***

103. 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*. 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 recorded copy of consultations for patients.

### ***Information***

104. 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*. A systematic review confirms that patients with cancer obtain benefit from accurate information, tailored to their diagnosis, stage and level of understanding.

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105. Consistent, but limited, evidence from observational studies suggests that much internet information on sarcoma is of poor quality, containing inaccuracies.
106. In a survey by the Sarcoma UK charity 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 given by doctors about their own situation.
107. 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.

### ***Psychosocial support***

108. Evidence from three systematic reviews, considered in the NICE guidance on Improving Supportive and Palliative Care for Adults with Cancer, 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.
109. 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***

- 110. 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.
- 111. Evidence relating travel to patient outcomes is inconclusive, because of the scarcity and heterogeneity of studies in this area.
- 112. 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.

**D. Measurement**

***Structure***

- 113. Provision of appropriate and adequate verbal and written information about the patients' diagnosis, proposed treatment options and sources of practical help.
- 114. Provision of training courses in communication skills for the relevant healthcare professionals.
- 115. Provision of self-help groups and other forms of support.
- 116. Provision of named key worker.

***Process***

- 117. Evidence that patients receive appropriate and timely information.
- 118. Evidence about the proportion of staff involved in patient care who have received formal communication training.

***Outcome***

119. Surveys of patient experiences of each component of patient-centred care.

**E. Resource implications**

120. [Resource implications will be available in the second consultation version of this document.]

## **Chapter 3 - Improving diagnosis of bone and extremity soft tissue sarcoma**

121. The over-riding 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.
122. 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.
123. 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***

124. For soft tissue sarcomas the principle problem in diagnosis is the large number of benign soft tissue tumours that cannot reliably be distinguished from malignant tumours (sarcomas) using clinical judgement.
125. The draft clinical guidelines (NICE Referral Guidelines for Suspected Cancer) 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 fails to address this need and this contributes to delay and adverse outcomes for those 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.

126. Patients with soft tissue lumps that do not meet the urgent referral criteria will be treated by GPs in the normal way.

### ***Bone Sarcomas***

127. 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 crucially on the recognition of an abnormal plain X-ray and failure to recognise abnormal X-rays frequently contributes to the diagnostic delay for patients with bone sarcomas. Plain X-ray films may demonstrate clear evidence or an abnormality strongly suspicious of a bone sarcoma but because of the rarity of primary malignant bone tumours, experience in interpreting abnormal X-rays is likely to be limited in non-specialist centres. 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***

128. Commissioners should consider publicity campaigns to increase public awareness of the signs and symptoms of sarcomas and the consequent need to attend their GP.

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129. It is recommended that commissioners ensure that GPs should be aware of and comply with the urgent referral criteria in the NICE Referral Guidelines for Suspected Cancer
130. Networks should ensure that GPs and hospital doctors are aware of the diagnostic pathways for patients with features suggestive of bone or soft tissue sarcoma.

### ***Referral pathways – extremity, trunk and head and neck soft tissue sarcomas***

131. In the absence of clear evidence of effective practice in the early diagnosis of STS this guidance proposes that a clearly defined network of diagnostic clinics, linked to sarcoma treatment centres (see Chapter 5) be established. Two possible models may achieve this:

EITHER:

1. Patients with a possible diagnosis of STS (as defined by the urgent referral criteria) would be seen at a diagnostic clinic that is part of a sarcoma treatment centre, within the two week wait.

OR:

2. Patients with a possible diagnosis of STS (as defined by the urgent referral criteria) would be seen at a specifically designated diagnostic clinic in their local cancer network, within the two week wait. This would purely be a diagnostic rather than a treatment clinic, and be clearly affiliated to one sarcoma MDT (see Chapter 5).

132. Each cancer network should designate such 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.
133. These diagnostic clinics (in either model) should undertake triple assessment including clinical assessment, imaging and biopsy of all

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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 suspected of having a STS should be rapidly referred on to a member of the sarcoma MDT for definitive treatment, as would any cases with equivocal images or biopsy.

134. A diagnostic clinic separate from a sarcoma treatment centre should have its staff trained and work audited by the Sarcoma MDT from the sarcoma treatment centre to which they are affiliated.
135. Appropriate imaging facilities should be available to comply with national access standards.
136. 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 a sarcoma treatment centre.
137. Commissioners and networks should work together to ensure that there are clear referral pathways from both primary and secondary care through to the closest diagnostic clinic and for proven sarcomas to a sarcoma treatment centre.

### ***Referral pathways – bone sarcomas***

138. All patients with a possible bone sarcoma should be referred directly to a bone tumour treatment centre (see Chapter 6) for diagnosis and management.
139. Appropriate imaging facilities should be available to comply with national access standards.

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140. The biopsy of patients with a possible bone sarcoma should only be carried out at a bone tumour treatment centre.
141. Patients with X-ray abnormalities which are most likely to be due to a secondary malignancy or are likely to be benign should be referred to the local orthopaedic service. Networks should consider formalising service provision for this latter group.

### ***Radiology review***

142. If a plain X-ray shows abnormalities which 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***

143. 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**

144. There will be a clear referral pathway from primary and secondary care to the appropriate sarcoma MDT and this will reduce delays in diagnosis.
145. 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.
146. Review by specialist pathologists of all tissue samples thought to be sarcoma will improve diagnostic accuracy.

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

### **C. Evidence**

#### ***Patient related delay in diagnosis***

148. 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.
149. In a Belgian study 47% of patients with soft tissue sarcomas showed delay of more than one 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 sarcomas waited for more than 6 months following the onset of symptoms before seeing a doctor. The average patient delay in a small UK study of patients with malignant bone or soft tissue tumours was 7.6 months.
150. The shortest patient delays were reported for patients with osteosarcoma; in 5 studies estimates ranged from 1 to 1.6 months. Patient delays were longer for those with Ewing's sarcoma; estimates from 5 studies ranged from 1.5 to 4 months.

#### ***Referral delay***

151. 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.
152. In a study of referral to a UK specialist soft tissue sarcoma unit, delay of more than three 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 patients' GP or local hospital took 7.5 months.
153. An unpublished observational study, using data from the Northern & 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.
154. In a recent UK survey of a group 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 a bone or soft tissue sarcoma was 4.85 in an American study.
155. 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 four papers. Longer doctor related diagnostic delays were reported for patients with Ewing's sarcoma of bone (1.25 to 7.75 months; 3 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 seven months in this group.
156. A large American study reported that 50% of patients with soft tissue sarcomas experienced a doctor-related diagnostic delay of two months or more and in 21% of cases delay was more than six months. Similarly in a Belgian study, doctor-related delay of more than one 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***

157. 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.
158. 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.
159. In a UK study of patients with soft tissue sarcoma, which 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 and referral delays tended to be longest for patients with benign tumours and shortest for those presenting with metastatic disease.
160. 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.
161. Two studies reported adverse outcomes in patients who had been misdiagnosed and managed inappropriately. In a series of patients with musculoskeletal tumours which had been misdiagnosed, 60% of cases required a more radical surgical procedure than would originally have been necessary due to 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 which 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***

162. 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.
163. Three observational studies of pre-referral MRI or 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***

164. A UK observational study found that 19% of bone tumours referred to a treatment service had been missed by both the clinician and 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.
165. There is consensus among radiologists working in the specialist areas that specialist review of the imaging in potential sarcoma patients reduces clinical error rates and delay in diagnosis.
166. 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 the interpretation between specialist and general radiologists.



***Biopsy***

167. 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. Three of these studies, one from the UK, one American and one Australian, 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***

168. 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**

169. The key factor in improving diagnosis and outcome for patients with sarcoma is referral for management to the sarcoma MDT before treatment. The patient pathway can be routinely audited and reviewed under the national cancer peer review system.

***Structure***

170. Provision of clear protocols and information on referral processes.
171. Provision of adequate staff and resources for effective functioning of all specialist sarcoma MDTs in order to assure compliance with waiting time requirements for diagnosis.
172. Provision of adequate access to appropriate imaging equipment and specialist radiologists.

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173. Sessional time and appropriate laboratory facilities for specialist histopathologists to attend specialist MDT meetings and provide a diagnostic service.

### ***Process***

174. Compliance with NHS targets for diagnosis and treatment relevant to sarcoma services.
175. The percentage of patients referred to the sarcoma MDT before the first definitive treatment in order to monitor the frequency of non-specialist treatment.
176. The percentage of sarcoma patients undergoing preoperative scan (MRI or CT) and staging investigations before first definitive treatment in order to monitor appropriate diagnostic management.
177. The percentage of sarcoma patients undergoing preoperative diagnostic biopsy (excluding retroperitoneal sarcomas) before first definitive treatment.

### ***Outcome***

178. Patient satisfaction.
179. Effect of diagnostic accuracy on patient outcomes.

## **E. Resource implications**

180. [Resource implications will be available in the second consultation version of this document.]

## Chapter 4 - Improving Pathology

181. 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 sub-classifications 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.
182. 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 appropriate experienced pathologist.
183. 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. Currently there is no established or funded laboratory for providing this service. These facilities require to be funded at one or more centres. Storage of tissue samples is important for future research in these rare tumours.
184. Gastro-intestinal stromal 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 pathologists with an interest in gastro-intestinal tumours. The diagnosis is based on tumour morphology, knowledge of tumour site and a panel of

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immunohistochemical markers, including CD117 (KIT). Demonstration of KIT expression is not sufficient to establish the diagnosis since it is not exclusive to this tumour.

185. 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 tumours.
186. 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 GI pathologist.
187. The Department of Health has made enrolment of NHS laboratories in accreditation schemes mandatory. There is almost exclusive use of Clinical Pathology Accreditation (UK) Limited 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**

188. All malignant bone tumours should either be first reported or reviewed by a SSP-bone. A SSP-bone is a pathologist who regularly reports bone tumours and these form a significant component of their workload and who successfully participates in the bone part of the Bone and Soft Tissue pathology EQA scheme.

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189. All soft tissue sarcomas should either be first reported or reviewed by an SSP-soft tissue. A SSP–soft tissue is a pathologist who regularly reports soft tissue tumours and these form a significant component of their workload and who successfully participates in the soft tissue part of the bone and soft tissue pathology EQA scheme.

190. All GISTs should be reported/reviewed by a 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.

191. All soft tissue tumours assessed in a diagnostic clinic (see Chapter 3) should have their pathology reviewed by:

EITHER:

- a SSP-soft tissue

OR

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

192. All malignant soft tissue tumours should be reported or reviewed by a SSP-soft tissue prior to management decisions by the sarcoma multidisciplinary team (MDT).

193. Pathology reports should include all the information required by the current Royal College of Pathologists' minimum dataset for soft tissue sarcomas once it is available.

194. There should be at least conditional accreditation for the laboratory in which the SSP and those with a specialist interest work.

195. There should be formal documented audit of the work of the SSPs and the nominated pathologists.

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196. The SSPs should have access to molecular pathology and/or cytogenetics facilities.
197. All sarcoma MDTs (see Chapter 5) must have at least one or, ideally two SSPs. All SSPs should work in collaboration with a colleague.
198. The additional work of reviewing cases by SSPs should be recognised in their job plan.
199. The Department of Health/Welsh Assembly Government should fund:
- A formal system for second opinions and review of difficult cases
  - Molecular pathology and cytogenetics facilities.
200. All pathology laboratories in centres treating bone or soft tissue sarcomas should have facilities for storing tissue for research (subject to the provisions of the Human Tissue Act).

### **B. Anticipated benefits**

201. The use of specialist pathologists will reduce the risk of errors in diagnosis in primary bone and soft tissue sarcomas.
202. Work undertaken in an accredited laboratory will help ensure high quality pathology services.
203. The establishment of a formal network between SSPs and nominated pathologists will help ensure accuracy in diagnosis and encourage training and education.
204. A national tissue resource will be created by a large tissue bank of sarcoma material for future research.

**C. Evidence**

205. There is consistent evidence that 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.
206. 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%.
207. Ten studies reported that the subtype of sarcoma was changed on expert review in between 16% and 39% of cases.
208. 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%.
209. One study reported a lower diagnostic error rate at musculoskeletal tumour treatment centres (13%) than at referring institutions (24%).
210. Central histopathological review as part of the European Osteosarcoma Intergroup clinical trial found 2% of the patients randomised to participate were in fact ineligible due to incorrect pathology.
211. 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

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relevant in the diagnosis of sarcomas that are difficult to distinguish histologically but where patient management depends on the diagnosis (for example: alveolar versus embryonal rhabdomyosarcoma; synovial sarcoma versus other spindle cell sarcomas or c-kit mutational analysis in GIST).

### **D. Measurement**

#### ***Structure***

- 212. Availability of specialist diagnostic laboratories.
- 213. Availability of specialist pathological review.

#### ***Process***

- 214. Time from GP referral to definitive diagnosis.
- 215. Time between biopsy and receipt of pathology report.
- 216. Attendance of SSP at MDT meetings.
- 217. Participation in an appropriate EQA Scheme.

#### ***Outcome***

- 218. Effect of diagnostic accuracy on patient outcomes.
- 219. Patient satisfaction.

### **E. Resource implications**

- 220. [Resource implications will be available in the second consultation version of this document.]



## Chapter 5 - Improving Treatment: Sarcoma Multidisciplinary Teams

221. Multidisciplinary teams (MDTs) have become the accepted way of delivering of 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.
222. 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 7-8 million.
223. The development and support for such a team needs 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.
224. 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.
225. The appropriate care of patients with less common soft tissue sarcomas is more complicated. More than one MDT will need to consider the management of the patient.

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226. These include head and neck, gastro-intestinal and uterine teams. Particularly complex cases may need referral to a more experienced MDT for advice.

### A. Recommendations

227. All patients with bone or soft tissue sarcomas should be managed by a sarcoma MDT.

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

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

230. There should be a nominated clinician (clinical lead) who takes responsibility for the service and this should be reflected in their job description.

231. All patients with a sarcoma must have their case discussed at an appropriately qualified MDT for that condition / site.

232. It is recommended that specific expertise of different MDTs should be made widely available so that cases can be referred expeditiously. Such expertise –which is not likely to be found everywhere – includes:

- Gynaecological sarcomas
- Head and neck sarcomas
- Gastro-intestinal stromal tumours (GIST)
- Central nervous system (CNS) sarcomas

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- Chest wall/intrathoracic sarcomas
- Skin sarcomas
- Fibromatosis

***Sarcoma MDT membership***

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

**Table 5. Core membership of a sarcoma MDT.**

<b>STAFF REQUIREMENTS</b>	<b>SPECIFICATION</b>
Specialist sarcoma surgeons	A minimum of two per MDT. These surgeons should have a major clinical interest in sarcomas i.e. spend >50% of their time in managing sarcomas.
Specialist sarcoma radiologist	At least two with a special interest in musculo-skeletal/ oncological imaging.
Specialist sarcoma pathologist	At least one and ideally two. In the event of a pathologist working in isolation, they should have close links with another SSP to allow double reporting and review of all cases (see Chapter 4).
Medical oncologist and a clinical oncologist	At least one of each. The oncologist/s should spend a significant portion of their time involved in the management of sarcomas.

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Paediatric oncologist	Specifically for bone sarcoma MDTs
Key worker	Sufficient to allocate a key worker for each patient – see Chapter 8 (but minimum of two).
Support staff	MDT co-ordinator and secretarial support.

Each MDT should in addition have an extended team with membership as shown in Table 6.

**Table 6. Membership of an extended sarcoma MDT.**

<b>STAFF REQUIREMENTS</b>	<b>SPECIFICATION</b>
Palliative care specialists	With an interest in sarcomas.
Specialist sarcoma physiotherapist	
Specialised rehabilitation team	Consisting of other relevant physiotherapists, occupational therapists, and orthotists with access to psychologists and artificial limb services.
Paediatric oncologist	A paediatric oncologist should be a member for appropriate cases.
Specialist nurses	Including palliative care, Macmillan nurses and appropriately trained ward staff.
Affiliated medical or clinical oncologist from linked cancer centre	Nominated by cancer network and approved by MDT
Affiliated diagnostic service clinicians	Nominated by cancer network

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	and approved by MDT
Other designated professionals	Including orthopaedic, thoracic, plastic, head and neck, gynaecological and GI surgeons.

234. 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***

235. The MDT should:

- Have weekly meetings at which all key members of the team should be present and at which all new cases should be discussed with documentation of attendance, decisions; other cases to be reviewed as necessary.
- Ensure that each patient has a written care/treatment plan which draws together the provision of all components of care.
- Ensure a key worker has been allocated to each patient.
- Co-operate in service development at a national and local level for patients with sarcomas.
- Ensure national standards for diagnosis and treatment are achieved.
- Have operational policies for the diagnosis and treatment of patients.
- Have documented arrangements for linking with other MDTs to ensure co-ordinated management of patients with sarcomas in a range of other anatomical sites i.e. head and neck, uterine, retroperitoneal and GIST (see Chapter 7).
- Comply with the information requirements of the National Cancer Dataset.
- Partake in a national audit program for sarcoma outcomes.
- Participate in national and international trials.
- Ensure audit and education of its referring hospitals and networks.

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- Ensure general practitioners are given prompt and full information about significant changes in their patients' illness or treatment.

### **B. Anticipated benefits**

236. All patients will be seen and assessed rapidly at a centre with appropriate expertise.
237. All aspects of the patients' management will be consistent and well co-ordinated, with appropriate use of specialist skills.
238. Co-ordinated specialist rehabilitation and supportive care will be provided.
239. National programmes will be supported and encouraged.
240. The sarcoma treatment centre will be the focus of continual improvements of the pathways of care for patients served by the centre.
241. More patients will be entered into clinical trials.

### **C. Evidence**

242. 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. Pre-treatment 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 soft tissue sarcoma***

243. 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 sarcomas at other anatomical sites. The UK study was the only one to adjust for differences in case mix in its analyses.

244. 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.
245. 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 59% of surgical resections in non-specialist treatment centres reported as 'wide' were found to contain residual disease on specialist pathological review.
246. Other differences between patterns of care provided by specialist sarcoma multidisciplinary teams and other treatment centres included

better conformity to clinical practice guidelines by multidisciplinary teams and greater use of preoperative imaging and biopsy.

***MDTs for bone sarcomas***

247. Evidence relating management 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 specialist MDT at the two supraregional bone tumour services or 20 UKCCSG paediatric oncology centres had improved 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 bone sarcomas.

***Hospital case volume and patient outcome***

248. There is consistent evidence, reviewed for example in *NICE Improving Outcomes in Colorectal Cancer*, that in complex or high risk surgery for cancer, case volume appears positively associated with improved patient outcomes.

249. Evidence about hospital case volume and outcome in patients with sarcoma was limited to two observational studies and a cohort study. Due to 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.

250. 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  $\geq 10$  cases. 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.



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251. A Dutch observational study compared the outcomes of patients with retroperitoneal soft tissue sarcoma in hospitals treating an average of less than one patient per year on average 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.
252. A large Canadian observational study of patients with extremity soft tissue sarcoma compared patient outcomes in three categories of hospital case volume: <2, 2-5 and >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***

253. Evidence that sarcoma MDTs have been established with the formal agreement of the cancer networks.
254. Sarcoma MDTs are staffed appropriately.
255. 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***

256. Evidence that the sarcoma MDT manage the minimum number of patients as defined in this guidance.
257. Every patient is discussed by a suitable MDT at the first opportunity after diagnosis.

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- 258. Participation by individual specialists in MDT meetings.
- 259. Audit and review by surgeons and histopathologists.
- 260. Operational policies for referral of patients to the centres.
- 261. Demonstration of links to appropriate specialist surgical expertise.

### ***Outcome***

- 262. Improvements of clinical outcomes.
- 263. Increase in clinical trial participation.
- 264. Patient satisfaction.

### **E. Resource implications**

- 265. [Resource implications will be available in the second consultation version of this document.]

## Chapter 6 - Improving Treatment: Bone Sarcomas

266. 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 the patients (median 21 years)
- the complexity of treatment
- requirements for rehabilitation

267. 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. Limb salvage can be achieved in 85% of patients, the remainder will need amputation.

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

269. 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.

270. 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 four to six weeks with daily attendances for treatment.

### **A. Recommendations**

271. There should be a formal relationship between the bone tumour treatment centre where surgical services are provided and the provider of oncology services characterised by common protocols, good communication, and well defined referral pathways.

#### ***Surgery***

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

273. 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 STS).

#### ***Chemotherapy***

274. The provider of chemotherapy services should:

- provide the facilities for intensive inpatient chemotherapy as described in the Manual of Quality Measures for Peer Review 2004.
- be EITHER:
  - a principal treatment centre for children and young people (likely to be a UKCCSG Centre or a Teenage Cancer Trust Unit)
- OR:
  - a cancer centre linked to a either a bone sarcoma MDT or one of the above units.
- have a nominated medical oncologist with a specific interest in chemotherapy for bone sarcoma

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- offer all patients with bone sarcomas entry into the relevant clinical trials
- provide facilities for long term follow-up for late effects of chemotherapy
- be guided by the bone sarcoma MDT on the treatment regimen.
- be a member of the extended bone sarcoma MDT

### ***Radiotherapy***

275. The provider of curative radiotherapy services should:

- be EITHER:
  - at a cancer centre linked to a either a bone sarcoma MDT
- OR:
  - be a principal treatment centre for children and young people.
- have a nominated clinical oncologist with a specific interest in radiation therapy for bone sarcoma.
- be guided by the bone sarcoma MDT on the treatment regimen.
- be a member of the extended bone sarcoma MDT

### ***Palliation***

276. 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.

## **B. Anticipated benefits**

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

278. 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.

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

## **C. Evidence**

### ***Specialist centres***

280. 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.
281. 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.
282. A small 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***

283. 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.

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284. A large unpublished 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 within paediatric oncology units was related to closer adherence to protocols in those units.

285. A systematic review which 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**

286. Membership of the sarcoma MDT as defined in Chapter 5.
287. Availability of common protocols and referral pathways with oncology providers.
288. Provision of and access to appropriate specialist surgical and non-surgical care.
289. Appropriate staff levels and training in the designated chemotherapy and radiotherapy centres.

#### **Process**

290. Compliance with nationally agreed waiting times.

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291. Audit of chemotherapy and radiotherapy regimens.

***Outcome***

292. Stage specific 5 year survival for patients with bone sarcoma.

293. Amputation rate in patients treated with curative intent.

294. Chemotherapy related toxic deaths.

295. Patient satisfaction.

**E. Resource implications**

296. [Resource implications will be available in the second consultation version of this document.]



## **Chapter 7 - Improving Treatment: Soft Tissue Sarcomas**

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

- Limb, limb girdle and truncal STS
- Abdominal and pelvic sarcomas
- Sarcomas requiring joint management

298. STS can rarely arise in other sites including the viscera and central nervous system.

### **Limb, limb girdle and truncal soft tissue sarcomas**

299. The commonest sites for STS 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.

#### **A. Recommendations**

300. The treatment (surgery, chemotherapy, radiotherapy) of all patients with limb, limb girdle and truncal STS should be decided by a properly constituted sarcoma MDT (see Chapter 5).

#### ***Surgery***

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

***Radiotherapy and chemotherapy***

302. The sarcoma MDT should recommend the treatment regimen.
303. Curative treatments should normally be delivered at the sarcoma treatment centre.
304. In specific situations curative treatment may be provided by a nominated clinical or medical oncologist with particular expertise who is not working at a sarcoma treatment centre. This person should be a member of the extended sarcoma MDT.
305. The preferred provider for palliative radiotherapy and chemotherapy services should be decided by the MDT in conjunction with the patient and agreed with local radiotherapy and chemotherapy providers.
306. 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 would be an extended team member of a sarcoma MDT (as defined in Chapter 5) and who would agree to give palliative treatments (chemotherapy or radiotherapy) according to protocols defined by the sarcoma MDT. These oncologists should be nominated by the cancer network and approved by the sarcoma MDT.

**B. Anticipated benefits**

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

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308. 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.
309. Outcomes and results can be recorded and expertise gained by the treatment team.

### C. Evidence

310. 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.
311. 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 1977-1984.
312. 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.

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

#### **D. Measurement**

##### ***Structure***

314. Membership of the sarcoma MDT as defined in Chapter 5.
315. Availability of common protocols and referral pathways with oncology providers.
316. Provision of and access to appropriate specialist surgical and non-surgical care.
317. Appropriate staff levels and training in the designated chemotherapy and radiotherapy centres.

##### ***Process***

318. Compliance with nationally agreed waiting times.
319. Audit of chemotherapy and radiotherapy regimens.

##### ***Outcome***

320. Stage specific 5 year survival, local control and complication rates.
321. Patient limb function and quality of life.

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322. Patient satisfaction.

**E. Resource implications**

323. [Resource implications will be available in the second consultation version of this document.]

## Abdominal and pelvic soft tissue sarcomas

324. Abdominal and pelvic STS pose particular challenges in treatment, especially in retroperitoneal sarcomas. 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.
325. 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.
326. GIST and uterine sarcomas are not included in this section because their management also needs to be considered by specific treatment teams.

### A. Recommendations

327. Patients with abdominal and pelvic STS should be referred to a sarcoma treatment centre where there is a team with a special expertise in managing these tumours.
328. NSCAG should consider funding of a number of designated specialist centres for the management of abdominal and pelvic soft tissue sarcomas.

### B. Anticipated benefits

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

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

331. 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**

332. 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 population based study of 143 patients with retroperitoneal sarcoma 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 resections were more likely at the higher volume hospitals, but overall survival was not related to case volume, in a case mix adjusted analysis.

333. In 24 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 18cm, which were predominantly high grade. Reports of 5 year overall survival varied between 19% and 63% for patients with localised primary disease and 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. Perioperative mortality, where reported, ranged from 0% to 9%.

334. 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. Due to 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 due changes in patient management practices and technologies over this time.

#### **D. Measurement**

##### ***Structure***

335. Evidence that there is a surgeon with specific expertise in these tumours who is a nominated contributor to the MDT and who attends meetings, when appropriate cases are to be discussed.

##### ***Process***

336. Evidence of participation by individual specialists at MDT meetings
337. Proportion of patients with these tumours referred to specialist sarcoma MDT with a specialist surgeon.

##### ***Outcome***

338. Stage specific 5 year survival, local control and complication rates.
339. Patient limb function and quality of life.
340. Patient satisfaction



**E. Resource implications.**

341. [Resource implications will be available in the second consultation version of this document.]

## Soft tissue sarcomas requiring joint management

342. STS 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
- Central nervous system (CNS) sarcomas
- Skin sarcomas
- Fibromatosis
- Gastro-intestinal stromal tumours (GIST).

343. Uterine sarcomas usually present with a pelvic mass which is sometimes asymptomatic. Often a diagnosis of sarcoma is only made post-operatively 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.

344. Head and neck sarcomas represent between 150-200 cases per year in England and Wales and between 10-15% 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 co-operation 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.
345. The management of chest wall and intrathoracic sarcomas requires a combination of skills available from a sarcoma MDT and a thoracic surgeon, often combined with plastic surgical reconstructive skills.
346. 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.
347. CNS sarcomas are rare and will generally be managed by a neurosurgical MDT.
348. Fibromatosis is a benign but infiltrative and destructive condition which simulates STS in its physical signs, 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.
349. Gastrointestinal stromal tumours (GISTs) are the commonest mesenchymal tumour to arise in the gastrointestinal (GI) tract. They represent approximately 1% of GI cancers and up to 5% of all soft tissue sarcomas. Although CT scanning is the standard staging investigation, 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 patients. Guidance on the use of imatinib in GIST was issued by NICE in 2004 (*NICE Technology Appraisal 86*:

*Imatinib for the treatment of unresectable and/or metastatic gastrointestinal stromal tumours*). This is a rapidly changing field. The role of imatinib is not yet fully understood and novel agents for treating GIST are likely to be available in the near future. This emphasises the importance of continued clinical research.

**A. Recommendations**

350. Patients with these sarcomas should be managed by the appropriate site specific MDT in conjunction with a sarcoma MDT.
351. The site specific MDT in conjunction with the sarcoma MDT should define the best team to deal with each patient. They should use specified care plans, taking into account currently available clinical trials.
352. Skin 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 another.
353. Patients with fibromatosis or other soft tissue tumours of borderline malignancy should be referred to a sarcoma MDT for diagnosis and management.
354. Commissioners should provide funding for imatinib following the recommendations made in the NICE Technology Appraisal.
355. Funding should be made available for clinical trials for the full evaluation of imatinib, other novel agents and the role of PET scanning in this condition.
356. Dietetic support should be available for patients who have undergone major abdominal surgery.

## **B. Anticipated benefits**

357. 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 both in terms of survival and local control.
358. There should be increased entry into relevant clinical trials.

## **C. Evidence**

### ***Joint management between site specific and sarcoma MDTs***

359. 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 decisions for these rare tumours require specialist knowledge.

### ***Imatinib***

360. NICE technology appraisal guidance 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. 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.
361. 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***

362. 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 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 US methods, with equivalent specificity.
363. 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.

**D. Measurement**

***Structure***

364. Referral protocols between the sarcoma MDT and the appropriate site specific specialist MDT, which clearly define the arrangements for joint discussion and management.
365. Appropriate specialist surgical expertise.
366. Specialist pathology review for patients diagnosed with GIST.
367. Provision of imatinib according to protocol.

***Process***

368. Proportion of patients with these tumours whose management has been jointly discussed at the sarcoma and site specific MDTs.
369. Audit of care and referral pathways.
370. Proportion of GIST patients receiving imatinib.

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***Outcome***

- 371. Patient satisfaction.
- 372. Stage specific 5 year survival, local control and complication rates.
- 373. Patient limb function and quality of life.

**E. Resource implications**

- 374. [Resource implications will be available in the second consultation version of this document.]

## Chapter 8 - Supportive and Palliative Care

375. The National Institute for Clinical Excellence (NICE) Guidance on Improving Supportive and Palliative Care for Adults with Cancer was published in March 2004. This chapter complements the guidance given, with specific reference to patient with sarcomas.

376. The five year survival of patients diagnosed with sarcoma is approximately 50% and there is a need for services to work together to provide co-ordinated 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, prosthetic limbs and for a wide spectrum of rehabilitation services. Support for sarcoma patients is a normal part of the sarcoma multidisciplinary team's (MDT) role. However, given that much of the treatment for patients with sarcoma is palliative, it is essential that the palliative care teams in the hospital are involved early and liaise directly with the community services.

377. This chapter describes four key components of care:

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

### The key worker

378. Key workers are individuals (usually a specialist nurse) who are familiar with sarcomas and their treatment and who can act as an advocate of the patients, facilitating the co-ordination of the diagnostic and treatment pathway, providing continuity and ensuring the patient knows



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how to access information and advice. They are a core member of the MDT and are involved prominently and personally in the patients' overall care. They liaise with health and social care teams, and professionals in the community, including the primary care team and palliative care when necessary.

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

### A. Recommendations

380. A key worker should be identified by the MDT for every patient with a sarcoma.

### B. Anticipated benefits

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

### C. Evidence

382. The NICE guidance on *Improving Supportive and Palliative Care for Adults with Cancer* considered interventions designed to improve the co-ordination of care. Two randomised controlled trials examined the co-ordination 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 health professionals.

**D. Measurement**

***Structure***

383. Identification of a key worker as part of the sarcoma MDT.

***Process***

384. Provision of a key worker to each patient for all stages of their treatment and care.

***Outcome***

385. Improved co-ordination of care.
386. Patient and carer satisfaction with the continuity of care.

**E. Resource implications**

387. [Resource implications will be available in the second consultation version of this document.]

## **Physiotherapy, occupational therapy and rehabilitation**

388. 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 sarcoma patients, especially teenagers and young adults is highly specialised. The role of the physiotherapist on the extended MDT enables rehabilitation to be provided in a timely and co-ordinated way. A range of other allied health professionals may be required at different stages in the patients' pathway and at a range of locations. Access to these services should be co-ordinated by the MDT.
389. 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.
390. Some patients will require specialist equipment such as compression hosiery, orthoses and environmental adaptations.
391. Clinical nurse specialists or key workers with appropriate experience and training can be extremely helpful in managing problems during the start of treatment including side effects and problems with nutrition, particularly in patients with GIST.

### **A. Recommendations**

392. A specialist sarcoma physiotherapist should be a member of the extended sarcoma MDT (see Chapter 5).
393. Ongoing rehabilitation and supportive care should be provided locally wherever possible. This should be co-ordinated by the therapist in liaison with the key worker.

## **B. Anticipated benefits**

394. Sarcoma patients would receive care from trained staff familiar with their condition.
395. Rehabilitation would be co-ordinated promoting a seamless service.

## **C. Evidence**

396. The NICE guidance on Improving Supportive and Palliative Care for Adults with Cancer reviews the evidence for the structure of effective rehabilitation services for adults with cancer.
397. 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.
398. 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 program. Another case report discussed the usefulness of a written plan during the rehabilitation of a young patient with Ewing's sarcoma.
399. 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. The review, limited to observational studies and expert opinion, concluded that the provision of a range of properly trained allied health professionals is essential during the rehabilitation of young people with cancer.

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400. The NICE guidance on Improving Supportive and Palliative Care for Adults with Cancer considers the evidence for the recommendation that allied health professionals should be part of cancer specific MDTs. Evidence was limited to professional guidance for allied health professionals which stressed the importance of multidisciplinary teamwork.

### **D. Measurement**

#### ***Structure***

401. Provision of adequately trained specialist physiotherapists and occupational therapists as part of the extended sarcoma MDT.
402. Evidence of established referral protocols.
403. Provision of necessary equipment.

#### ***Process***

404. Evidence that patients receive adequate input from specialist therapists.
405. Evidence of adequate arrangements for long-term care provision.

#### ***Outcome***

406. Patient satisfaction.
407. Quality of life and functional status.

### **E. Resource implications**

408. [Resource implications will be available in the second consultation version of this document.]

## Orthotic and prosthetic appliance provision

409. 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 life-long 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.
410. 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, inability to wear with some clothing and appearance.
411. Presently Disablement Service Centres (DSC) 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 in 2003. These centres have the necessary expertise and facilities to cope with all the intricacies of comprehensive prosthetic/amputee rehabilitation.

### A. Recommendations

412. Rapid, easy access should be provided to appropriate orthotic and prosthetic services.
413. The sarcoma MDT should establish formal links to a centre(s) matching the PARC template, including that patients should be referred for pre-amputation assessment.

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

### **B. Anticipated Benefits**

415. Improved functioning and quality of life.
416. Improved integration into society.

### **C. Evidence**

#### ***Satisfaction with prosthetics services***

417. 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.
418. 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 Disablement Services Centres, suggests that examples of good service provision exist.

#### ***Specialised rehabilitation service***

419. Expert opinion held that many Disablement Services Centres 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 Disablement Services Centres meeting the specifications of a Tertiary Referral Prosthetic and Amputee Rehabilitation Centre (PARC), as defined in the British Society of Rehabilitation Medicine (BSRM) standards and guidelines for amputee and prosthetic rehabilitation.

***Prosthetics for leisure activities***

420. A cohort study, reporting the incidence and aetiology of limb amputation in the UK (2003-2004), found that people who lose a limb due to 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 or leisure pursuits.

**D. Measurement**

***Structure***

421. Links with PARCs.
422. Provision of adequately trained specialist orthotists and prosthetists.
423. Provision of special activity limbs.

***Process***

424. Pre-op referral to PARC for assessment.
425. Evidence of long-term care provision.

***Outcome***

426. Patient and carer satisfaction with orthotic and prosthetic appliance provision.

**E. Resource implications**

427. [Resource implications will be available in the second consultation version of this document.]



## Specialist palliative care

428. Palliative care is essentially a community service and needs to be provided locally for sarcoma patients as required. There is however also a need for specialist palliative care input for some patients and specialist palliative care teams should be based in the unit in which the sarcoma MDT operates. The palliative care team will liaise with the local hospital and community teams as necessary.

### A. Recommendations

429. Palliative care specialists, with an interest in sarcomas, should be a member of the extended sarcoma MDTs.

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

431. Commissioners should ensure that sarcoma patients have easy and timely access to appropriate palliative and specialist pain management services.

### B. Anticipated benefits

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

433. 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.

434. 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 the support for close family members when it becomes clear that death is inevitable.

### **C. Evidence**

435. No evidence was identified on the effectiveness of palliative care teams with an interest in sarcoma. The NICE guidance on *Improving Supportive and Palliative Care for Adults with Cancer* reviews evidence for the configuration of palliative care services:

#### ***Palliative care specialists with an interest in sarcoma***

436. 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 palliative care between the MDT and local services***

437. Evidence from systematic reviews suggests that 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***

438. 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.

#### ***Co-ordination between hospital and community-based teams***

439. Two randomised controlled trials reported that employment of a nurse co-ordinator, 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***

- 440. Palliative care teams to support patients at home or in hospices.
- 441. Telephone support, advice and information services for patients and their carers.
- 442. Bereavement counselling for family members and carers as appropriate.

### ***Process***

- 443. Attendance of the palliative care specialist at the sarcoma MDT.
- 444. Evidence that providers elicit information about patient preferences about place of death and their views about medical intervention in the terminal phases of illness.
- 445. Regular systematic psychological assessment at key points and access to appropriate psychological support.

### ***Outcome***

- 446. Patients' experience of pain and satisfaction with pain control during treatment.
- 447. Symptom control and quality of life.
- 448. Patient and carer satisfaction with the services provided during the patient's final month of life.

## **E. Resource implications**

- 449. [Resource implications will be available in the second consultation version of this document.]

## Chapter 9 - Follow-up of patients

450. 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 so thus will never need following up. It is also time-consuming and expensive and may also produce anxiety rather than reassurance for patients.
451. For patients with GIST there is a much higher risk of recurrence which is likely to be intra-abdominal.
452. For patients enrolled in sarcoma 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. (eg. PET or CT for GIST).
453. Long-term follow-up will be needed for many patients, especially those who have received a prosthetic replacement or had a childhood cancer, because of the risk of late complications.

**A. Recommendations**

454. Commissioners should ensure that follow-up protocols are clearly defined and nationally agreed for each tumour type / location.
455. Resources should be made available for regular imaging of patients at high risk of recurrence (as defined in an agreed protocol).

**B. Anticipated benefits**

456. Clearly defined protocols for follow-up will improve the consistency and equity of care for these patients and the appropriate use of resources.
457. Early detection of recurrent disease.

**C. Evidence**

***Current situation***

458. A review article identified eleven 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.
459. 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 NCRI study in the UK produced very similar findings.

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

***Effectiveness of follow-up strategies***

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

462. 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. 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.

463. 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.

464. The effectiveness of routine follow-up testing was also considered in an unpublished 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.

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465. Evidence on the acceptability of follow-up to patients 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.

### ***Late effects of treatment***

466. 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. 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***

467. Nationally agreed protocols for follow-up.

### ***Process***

468. Audit of follow-up practices and the timeliness and appropriateness of investigation.

### ***Outcome***

469. Patient/carer satisfaction.

## **E. Resource Implications**

470. [Resource implications will be available in the second consultation version of this document.]

## Chapter 10 - Improving Knowledge

471. Sarcomas are rare tumours and reliable data on management and outcomes is largely limited to individual units that manage the conditions.
472. 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.
473. 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.
474. Implementation of the nationally agreed dataset for sarcoma as a subset of the National Cancer Dataset 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.
475. Training and continuing professional development (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*

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



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477. The data collected should be agreed nationally and should be based on the sarcoma subset of the National Cancer Dataset. Cancer networks should ensure that a complete dataset exists for all patients managed within their network.
478. 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***

479. Audit should be carried out of all elements of the referral and management pathway including standards for referral, investigation and management.
480. Pathology audit should continue to be undertaken by the existing EQA scheme and networks should ensure that only specialist sarcoma pathologists who comply with this scheme report on sarcomas.
481. 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 Steering group should be asked to provide guidance on multicentre audits.
482. Audits of outcome including patient satisfaction should be carried out by networks and sarcoma MDTs.
483. The results of audits should be widely available to both clinicians within referring units, networks and to the public.

***Training***

484. 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.
485. Appropriate training posts should be made available nationally to train and recruit surgeons, pathologists, radiologists and oncologists with appropriate expertise in sarcoma care.
486. MDTs should ensure that they regularly provide updates for members of the extended MDT.

***Research***

487. 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 configuration of services. It is therefore important that health service commissioners should support the well-designed clinical trials within the National Cancer Research Network (NCRN) portfolio.
488. 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.
489. Commissioners should ensure that NCRN adopted clinical trials for patients with sarcomas are supported locally.
490. All sarcoma MDTs should aim to maximise entry into trials and should work with the local NCRN to ensure this happens.
491. The possibility of entry into an appropriate trial should be discussed with every patient who fits the inclusion criteria. Such patients should be

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given accurate and accessible information to inform their decision about whether to participate in the trial.

492. 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.

493. 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***

494. 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.

495. 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***

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

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

**Research**

498. 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**

499. The rarity of sarcomas means that data needs 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.
500. Evidence from three observational studies, 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.

**Clinical trials and protocol-based care**

501. 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.

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502. 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*). 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*

503. Network-wide information systems that capture standard data of all sarcoma patients according to the national cancer dataset.
504. Availability of support to collect data and enable it to be shared within and between networks.

##### *Process*

505. Evidence that all patients' data is collected in accordance with national protocols.
506. Evidence that pathology data is collected and is used to assess the pathology department's performance as part of the EQA system.

#### AUDIT

##### *Structure*

507. Availability of support to carry out multicentre audits.
508. Agreements between cancer networks and sarcoma MDTs about the audit tool and frequency of multicentre audits.
509. Support for patient groups for carrying out patient satisfaction audits on behalf of sarcoma patients.

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### ***Process***

- 510. Proportion of sarcoma MDTs participating in regular audit and multi site audits.
- 511. Evidence of feedback and the development of action plans to referring clinicians following audit.

## **CLINICAL TRIALS**

### ***Structure***

- 512. Network-wide information systems that allow clinicians to identify trials for which specific patients might be eligible.
- 513. Availability of support for clinical trials.
- 514. Availability of continued support for patients who have been successfully treated with products used in clinical trials.

### ***Process***

- 515. Evidence of regular discussion of participation in clinical trials at MDT meetings.

### ***Outcome***

- 516. Proportion of patients with each type of sarcoma entered into trials.

## **E. Resource implications**

- 517. [Resource implications will be available in the second consultation version of this document.]