## SCHEDULE 2 – THE SERVICES

### A. Service Specifications

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| **Service Specification No.** | |
| **Service** | Sarcoma |
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| **Provider Lead** | Jeremy Whelan |
| **Period** | 12 months |
| **Date of Review** | |

### 1. Population Needs

**1.1 National/local context and evidence base**

This document provides a national specification for the care of all patients with sarcoma. It is underpinned by a single principle that all patients with a sarcoma will be referred to a specialised sarcoma service.

This model of sarcoma service aims to deliver high quality care to patients with suspected sarcoma from diagnosis through to specialist treatment and follow up. It defines the pathways for all patients with a sarcoma.

Sarcoma care is characterised by the need for complex multimodality assessment and therapy that involves pre-intervention evaluation - clinical, radiological, pathological and multidisciplinary; complex surgery for resection and reconstruction aimed to preserve function; further pathological assessment including margin description; complex radiotherapy; intensive chemotherapy; unique rehabilitation needs; and palliative care.

Sarcomas are a multitude of diseases with many unique and some shared characteristics. Their overall rarity and the multiplicity of presentation features determine that they are unfamiliar in routine clinical care. Management is frequently complex and pathways of care are complicated and individualised.

Sarcomas are rare cancers that develop in the muscle, bone, nerves, cartilage, tendons,

blood vessels and the fatty and fibrous tissues and fall into three main types:
i) Bone sarcoma
ii) Soft tissue sarcoma
iii) Gastrointestinal stromal sarcomas (GIST)

There are a number of associated benign or so-called 'borderline' conditions that appropriately fall in the remit of sarcoma services. The commonest example is fibromatosis.

The most common sites for both soft tissue and bone sarcomas are within the extremities (23% of soft tissue sarcomas and 52% of bone sarcomas). However sarcomas arise in all parts of the body producing specific clinical scenarios and challenges to management.

Survival at 5 years for soft tissue and bone sarcomas is 55% (data for 2006-2010) and 56% (2001-2005) respectively but there is considerable variation within this determined by histological subtype, stage and age .

**1.1.2 Key facts and evidence base**

There are around 100 different sub types of sarcoma and about 3,800 new cases of sarcoma are diagnosed each year in the UK which makes up approximately 1% of all cancer diagnosis.

- 3,300 people are diagnosed with a soft tissue sarcoma (including GIST).
- 500 people are diagnosed with a bone sarcoma
- 10 people every day are diagnosed with a sarcoma in the UK
- Sarcomas make up 15% of all childhood cancers (0-14 years)
- Sarcomas make up 11% of all cancer diagnosis in teenagers and young people (15-25 years)

In general, patients with a soft tissue sarcoma or bone sarcoma tend to be younger than the majority of cancer patients. 57% of soft tissue sarcomas affect those under 65 years and about a quarter of all bone sarcomas occur before the age of 30 years.

Services for sarcoma in England are based on:-

- Improving Outcomes for People with Sarcoma (NICE, March 2006), and subsequent supplementary guidance and measures which have underpinned peer review of sarcoma services (Manual for Cancer Services, Sarcoma Measures version 1, January 2014).
- Improving Outcomes in Children and Young People with Cancer (NICE August 2005)
- NICE Quality Standard for Sarcoma (NICE January 2015).
- Cancer waiting times
- Other national standards

See appendix 1 for links to the above

**1.1.3 Health needs, epidemiology, incidence,**

**Bone sarcoma**

Based on data from 1996-2010, the age-standardised incidence of bone sarcoma was

constant at around 7.9 per million.

Age specific incidence rates for bone sarcoma are bi-modal, with incidence peaks observed in teenagers and young adults, as well as the elderly. Bone sarcoma age specific incidence rates are significantly higher in males than females over 15 years of age. Rates in males exceed those of females by a ratio of 1.7:1 in those aged 15 to 19 years and by a ratio of 1.6:1 in those aged 55 years and over.

The four most common types of bone sarcoma are:

- Chondrosarcoma (37% of all bone sarcoma diagnosis)
- Osteosarcoma (30% of all bone sarcoma diagnosis)
- Ewings sarcoma (14% of all bone sarcoma diagnosis)
- Chordoma (6% of all bone sarcoma diagnosis)

The remainder are a variety of other sarcomas. Benign bone tumours including aneurysmal bone cysts and giant cell tumours may mimic malignant tumours and require the same diagnostic and surgical expertise.

Three main subtypes of bone sarcoma account for 75% of all cases. With complex intensive combined modality treatment about two thirds of patients will be cured. Incidence of chondrosarcoma increases with age and the main modality of treatment is surgery.

Most patients with bone tumours present with pain (particularly at night or non-mechanical) and/or swelling. Pathological fracture is a less common presentation feature. It is not uncommon for symptoms to have been present for some time before medical attention is sought or further investigations/referrals are initiated. Most occur in the bones of the extremity; however craniofacial , pelvic and axial skeletal tumours all occur and present specific management challenges.

The treatment required is dependent on diagnosis and stage of the disease. Treatment modalities used include surgical treatment and/or further treatment such as chemotherapy and radiotherapy. Surgical treatment involves excision of the tumour followed by surgical reconstruction. Surgical reconstruction can be:

- endoprosthetic replacement
- biological reconstruction
- composite reconstruction

Limb salvage may not always be possible, dependent on the site of the primary, and an amputation may still be required in up to 10% of cases.

The prognosis is dependent on the diagnosis, stage of the disease and response to chemotherapy.

Bone sarcomas account for about 6% of cancer in 0-14 year olds (average 55 cases per

year in UK) and 5% in 15-24 year olds (average 103 cases per year in UK). About a quarter of all bone sarcomas occur before the age of 30 years.

**Soft tissue sarcoma**

Based on data from 1996-2010, soft tissue sarcoma incidence rates increased significantly from 39 per million to 45 per million (this increase may reflect improved diagnostic techniques and reporting rather than a true increase in incidence).

The incidence of soft tissue sarcomas increases significantly with increasing age. The age specific incidence rate is highest in males aged 85 years and over where it reaches 230 per million and exceeds the rate for females by a ratio of 1.9:1.

Age specific incidence rates for soft tissue sarcomas in females aged 45 to 59 years are slightly higher than those in males, due to the incidence of gynaecological sarcomas.

In contrast to commoner cancers, an important proportion of soft tissue sarcomas affect younger people, with 57% of soft tissue sarcoma affect those under 65 years.

In their early stages, soft-tissue sarcomas usually do not cause symptoms because soft tissue is relatively elastic. Tumours can grow large, pushing aside normal tissue, before they are felt or cause problems. Often the first sign is a painless lump or swelling; however, as the tumour grows, the lump may cause pain as it presses against nearby soft tissues and nerves.

The most commonly occurring soft tissue sarcomas are extremity and trunk tumours and make up 60% of all adult soft tissue sarcomas. Patients present with a painless lump and it can be difficult to differentiate a benign from a malignant mass without specialist investigation.

Soft tissue sarcomas of the retroperitoneum and abdomino-pelvic cavities provide particular challenges in diagnosis as symptoms are usually late and non-specific (e.g. indigestion, constipation, menstrual cramps), and are often interpreted by non-specialist doctors as diagnostic of other problems.

Soft tissue sarcomas are an important cancer of children and young people, accounting for about 6% of cancer in 0-14 year olds (average 90 cases per year in UK) and 4% in 15-24 year olds (average 80 cases per year in UK). Rhabdomysarcoma, a form of soft tissue sarcoma arising from muscle progenitor cells, accounts for 31% of children diagnosed with sarcoma with a peak incidence at age 3 years but becoming less frequent after 6 years.

### 1.1.4 Inequalities

There are a number of inequalities experienced by sarcoma patients. Because of the rarity of sarcomas, patients may receive inappropriate treatment by non-skilled practitioners before the diagnosis has been made and the need for specialised care has been recognised. Depending on where sarcoma patients live in the England, there is variability in availability of specialist care, and in access to that care. A significant number of patients are treated outside of specialist sarcoma services.

Social deprivation has an impact on presentation and outcomes in sarcoma. The route to

presentation for sarcomas is more likely to be through GP referral or emergency presentation for patients in the most deprived areas. Furthermore, survival for those from more deprived areas is lower after presentation through an emergency route, and even for those referred by a GP from areas of the greatest deprivation. Moreover there is some evidence that the incidence of osteosarcoma may be increased in areas of lower deprivation.

The treatment of sarcomas in the elderly is often challenging, given that standard chemotherapeutic approaches are often inappropriate. Older patients with soft tissue sarcoma may have more advanced stage at diagnosis, receive less aggressive treatment and be less likely to participate in clinical trials.

**1.1.5 Scale and pattern of service**

The scale of sarcoma services provides cover across all Regions in England. The pattern of service is based on sarcoma multi-disciplinary teams in designated Specialist Sarcoma Centres hosting either a soft tissue sarcoma MDT (10 centres) or a combined bone and soft tissue sarcoma MDT (5 centres) and delivering defined, comprehensive sarcoma services serving a population within a defined Sarcoma Network.

A soft tissue sarcoma (STS) MDT must meet minimum criteria and manage the care of at least 100 new patients with soft tissue sarcoma per year.

If a sarcoma MDT manages the care of patients with both bone sarcoma and soft tissue sarcoma, it must manage the care of at least 50 new patients with bone sarcoma per year and at least 100 new patients with soft tissue sarcoma per year.

Each Sarcoma Network has a Sarcoma Advisory Group (SAG).

Local Sarcoma Units that have been designated by the SAG deliver defined elements of care for sarcoma patients more locally and work with the Sarcoma MDT at the Specialist Sarcoma Centre.

## 2. Outcomes

### 2.1 NHS Outcomes Framework Domains

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|---|---|---|
| **Domain 1** | **Preventing people from dying prematurely** | √ |
| **Domain 2** | **Enhancing quality of life for people with long-term conditions** | √ |
| **Domain 3** | **Helping people to recover from episodes of ill-health or following injury** | √ |
| **Domain 4** | **Ensuring people have a positive experience of care** | √ |
| **Domain 5** | **Treating and caring for people in safe environment** | √ |

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| | **and protecting them from avoidable harm** | |

**The sarcoma service metrics linked to the aim and objectives set out in section 3 of this specification are listed in Appendix 2.**

## 3. Scope

### 3.1 Aim and objectives

#### 3.1.1 Aim

The aim of the service is to improve outcomes for all patients with sarcoma by ensuring that all patients will be referred to specialised sarcoma services and that all patients have access to the highest quality care regardless of where they live or the location of the tumour. This includes access to surgical teams with appropriate sarcoma-specific and anatomic site knowledge as well as access to specialist paediatric, clinical and medical oncologists who have a particular interest and knowledge in sarcoma management with access to systemic treatment and radiotherapy; these clinicians must be supported by nursing and AHP staff who have also appropriate expertise.

Specialist Sarcoma Centre multidisciplinary teams are responsible for assessment, diagnosis and treatment including surgical management, oncology and radiotherapy for soft tissue sarcoma and bone sarcoma.

Pathways describing diagnosis, treatment and follow up supplemented by practice guidelines must be used to ensure consistent and equitable care.

Specialist Sarcoma Centres work according to prescribed pathways and guidelines with other service providers for the delivery of elements of care that may be appropriately undertaken outside of the Sarcoma Centre. In particular, practitioners designated by the SAG in conjunction with the Sarcoma MDT will assist in the delivery of elements of care (e.g. diagnosis, chemotherapy or radiotherapy), at Local Sarcoma Units.

#### 3.1.2 Objectives

All patients with sarcoma will be referred to a specialist sarcoma service. The sarcoma service objectives are:-

- To provide a comprehensive service for all patients with sarcoma within a SAG-led network.
- To provide expert accurate diagnosis and staging of bone and soft tissue sarcoma utilising the most up-to-date validated diagnostic tools.

- To have pathways for referral and diagnosis in place for people with suspected sarcoma.
- To ensure all patients with confirmed bone or soft tissue sarcomas have access to expert care and management provided by trained staff and receive care in accordance with the most up-to-date network agreed clinical protocols.
- For adults, children and young people with bone sarcoma and adults and young people with soft tissue sarcoma to have their care plan confirmed by a sarcoma MDT and treatment delivered by services designated by the SAG.
- To ensure collaborative working between paediatric, TYA and sarcoma MDTs.
- To support patients through an allocated key worker with specialist knowledge of sarcomas and their treatments.
- To ensure that all patients receive a written care plan.
- To provide chemotherapy, radiotherapy and surgery in line with national guidelines, evidence based practice and network policy and delivered by designated practitioners who are core or designated members of a Sarcoma MDT.
- To ensure optimal physical functioning and quality of life for patients with sarcoma through rehabilitation and monitoring.
- To work with Local Sarcoma Units to manage patients with sarcomas in line with national guidelines, evidence based practice and network policy.
- To ensure that patients have opportunities to participate in research
- To publish information about shared pathways, activity and patient outcomes, including information on site specific sarcomas.
- To provide high quality information for patients, families and carers in appropriate and accessible formats and mediums.
- To ensure that there is involvement of service users and carers in service development and review.
- To ensure there is a commitment to continued service improvement and to improved outcomes for patients with sarcoma through research and audit.
- For all sarcoma MDTs to present process and outcome data to the annual national audit meeting.
- To be compliant with National Guidance and standards including NICE IOG and standards.
- To ensure compliance with Care Quality Commission regulations.

**3.2 Service description/care pathway**

This section provides a description of the service model and care pathways for sarcoma services. It outlines the structural elements of sarcoma services, the people involved in delivering treatment, the technology needed to delivery sarcoma services and the processes involved linked to the care pathway.

**3.2.1 Service model and pathways**

As outlined in section 1.1.5 (scale and pattern of service), sarcoma centres host **sarcoma MDTs** for patients within a **sarcoma network.** Sarcoma networks have a **SAG** and **local sarcoma units** deliver elements of care for patients more locally.

**Sarcoma Networks**

A network for sarcoma services must include a Specialist Sarcoma Centre hosting an MDT and with network pathways to ensure a comprehensive service is available for patients with any type of sarcoma. This will also include elements of care delivered beyond the network for example for very rare sarcomas requiring very specialised care including but not confined to bone sarcomas.

The network pathways are the responsibility of the network SAG and are developed in conjunction with the sarcoma MDT and will include arrangements with other centres and Local Sarcoma Units within the network for the delivery of diagnostic or treatment services outside the centre by designated members of the sarcoma MDT.

Sarcoma Networks must have clear communication and dissemination processes between primary and secondary care and the Specialist Sarcoma Centres.

The sarcoma networks are responsible for improving access to specialist sarcoma services, improved operating consistency, improved outcomes and improved productivity. The network must be hosted by a Specialist Sarcoma Centre and will have clear links to the national Sarcoma Clinical Reference Group most often through the appointed clinician Network Chair of the Sarcoma Advisory Group (SAG). Networks must be underpinned by governance frameworks to ensure accountability, management of risk and delivery.

**Sarcoma Advisory Groups**

Sarcoma Networks will convene a SAG that must as a minimum fulfil section 14-1c of the Sarcoma Measures. The SAG provides the primary source of clinical opinion for sarcoma within a Sarcoma Network.

The SAG is responsible for reporting on the clinical elements of the service specification and the operational service development of the network to the the Lead Provider Trust, Local Sarcoma Unitsand local specialist commissioners. The SAG is also responsible for the establishment and operation of appropriate pathways, the arrangements for Specialist Sarcoma Centres working with Local Sarcoma Units, designation of practitioners, audit of services provided within the Network and sarcoma clinical trials within the Network.

Adjoining SAGs must work together to ensure that pathways for patients near borders who may be served by either of two or more Network services, are clear and not confusing.

SAGs must participate in a sarcoma national audit programme addressing service quality and outcomes.

**Specialist Sarcoma Centres**

Specialised Sarcoma Centres will host the MDT who will provide diagnostic, treatment and follow up services in conjunction with its respective Local Sarcoma Units.

Sarcoma MDTs will ensure up to date information about their shared pathways, activity and patient outcomes, including information on site-specific sarcomas is publically available.

Diagnosis of suspected primary bone tumours and the surgical treatment of primary bone tumours is delivered across England by the five designated centres which provide cover across all regions in England for the national caseload and which host or co-host combined bone and soft tissue sarcoma MDTs.

**Local Sarcoma Unit**
Trusts providing sarcoma care in conjunction with the Specialist Sarcoma Centres will be defined by the SAG. The principle underlying these arrangements is the delivery of safe, effective specialist care as locally as possible rather than local care as safely as possible, while recognising the diversity of clinical need inherent in a sarcoma population.

Services outside the Specialist Sarcoma Centre can include diagnostic services for suspected soft tissue sarcoma, primary surgical resection by designated members of the sarcoma MDT of some visceral sarcomas (e.g. uterine sarcoma) and delivery of some chemotherapy and radiotherapy, by designated members of the Sarcoma MDT, in accordance with recommendations from the Sarcoma MDT. Specialist Sarcoma Centres must ensure that services available at designated Local Sarcoma Units and details of designated practitioners are published with information about pathways.

**Sarcoma MDT**
The principal role of a Sarcoma MDT is to determine a care plan for all patients with bone and soft tissue sarcoma and to be responsible for its delivery either by members based at the Specialist Sarcoma Centre or by designated MDT members working at Local Sarcoma Units. The MDT will be constituted and organised in accordance with the Sarcoma Measures.

Sarcoma MDTs must publish information about their shared pathways, activity and patient outcomes, including information on site-specific sarcomas.

Sarcoma MDTs must include key workers so that all people with a sarcoma can be supported by an allocated key worker with specialist knowledge of sarcomas and their treatment.

The development of experience and expertise required of individual clinicians and other staff to treat patients with sarcoma is a major investment for all Specialist Sarcoma Centres. Each Centre must ensure that a programme of training and a succession planning strategy is in place.

Designated Local Sarcoma Units and Designated Practitioners deliver defined areas of care in conjunction with the sarcoma MDT.

These include

- Practitioners involved in the diagnosis of suspected soft tissue sarcomas presenting to rapid access diagnostic clinics linked to sarcoma MDTs.

- Practitioners working in other MDTs to which sarcomas may present. These include: breast, head and neck, brain and CNS, urology, gynaecological, urology, gastro-intestinal, lung, skin MDTs. Management of sarcomas arising in these sites must be in accordance with pathways agreed and monitored by the MDT in conjunction with the SAG.
- Practitioners delivering chemotherapy and radiotherapy services when this is deemed appropriate by the Sarcoma MDT to be undertaken outside the Specialist Sarcoma Centre.

**Service users**
All patients with suspected or confirmed sarcoma must have access to appropriate diagnostic, treatment and support services. These services must be patient centred and must respond to patient and carer feedback.

Sarcoma patient experience is reported in the National Cancer Patient Experience Survey, and highlights key areas where patient experience can be improved. Participation of service users in the operation and improvement of sarcoma services must be supported at every level. Service user representatives should participate in SAG activities.

**Diagnostic services**
Diagnosis of sarcoma is based on expert interpretation of imaging and histological material. Details depend on the anatomical location of the primary tumour. Extremity soft tissue sarcomas present as a small proportion of 'lumps and bumps' and clinical concerns may be initially investigated using ultrasound or MRI. Correlation of imaging features with histology is essential in sarcoma diagnosis, especially for bone sarcomas. An increasing number of sarcomas have unique chromosomal translocations so that molecular analysis is an increasingly important component of diagnosis, offering diagnostic, prognostic and therapeutic direction. Inappropriate biopsies or unplanned intralesional operations may have significant consequencesincluding delay and reduced chance of local tumour control.

**Surgical Services**
Surgery holds a position of critical importance for the majority of newly diagnosed sarcomas. For many, it is curative by itself, for others it is a vital part of multimodality therapy given with the intent of cure. For others, repeated and often complex surgical operations may play a valuable part in palliation.

The costs of inappropriate or inadequate surgery can be very high for patients with sarcoma, resulting in excess lifelong morbidity and reduced survival chances. Sarcoma services must be structured and managed to reduce the number of unplanned excisions and to ensure that all resections of sarcomas are undertaken by surgeons who are core or designated members of the Sarcoma MDT (see appendix 3). Audit of procedures undertaken by designated and non-designated surgical practitioners will be undertaken routinely using national data sets.

**Radiotherapy services**
Radiotherapy is a crucial part of primary local therapy for soft tissue and bone sarcomas, and also of palliation of sarcoma patients. Treatment planning and delivery to a high standard is essential for best outcomes. The rarity of sarcomas means that high

standards cannot be assumed. Successful outcomes rely on the experience of the treating clinical oncologist and their radiotherapy centre in sarcoma radiotherapy. Facilities must be provided according to the Manual for Cancer Services and the following structural/organisational points below must be met:

**Structural/organisational**

- **Treatment decisions** for delivering radiotherapy; these must be made within the Sarcoma MDT
- **Mould room services:**: the delivery of high quality radiotherapy, particularly for limb sarcomas, requires high quality immobilization, provided by mould room services. This is essential for any centre delivering sarcoma radiotherapy, whether a Specialised Sarcoma Centre, or a designated practitioner, and in particular for those delivering intensity modulated radiotherapy (IMRT).
- **Planning experience:** clinicians planning radiotherapy for sarcomas must have sufficient site-specialist planning experience and cross cover.
- **Referral pathways:** there must be effective communication between surgeons and oncologists (clinical and medical) to ensure the smooth transfer of patients between services in order to deliver the components of treatment within optimal timing and to minimise un-necessary delays. Clear pathways must be in place to ensure timely referrals of patients.
- **Bone sarcomas:** given the particular rarity of bone sarcomas, and the complexity of the required radiotherapy planning, radical radiotherapy will only very infrequently be delivered outside Specialised Sarcoma Centres.
- **Clinical trials:** Specialised Sarcoma Centres must participate in relevant clinical trials involving radiotherapy for sarcoma patients.
- **Specialised Sarcoma Centres:** will undertake regular audits of radiotherapy for sarcomas treated both at the centre and by designated practitioners.

**People**

- **Clinical oncologists:** are core members of the Sarcoma MDT. Most patients will be treated by site-specilaist clinical oncologists within the sarcoma MDT of a Specialised Sarcoma Centre. Radical and palliative treatments will be delivered.
- **Designated practitioners:** are nominated clinical oncologists at a radiotherapy centre agreed by their host Trust Clinical Director or equivalent and approved by the SAG. Referrals to designated practitioners will be from a clinical oncologist who is a core member of the Sarcoma MDT. (See appendix 3)

It is recognised that patients may live at some distance from a Specialised Sarcoma Centre, and it may not be possible for them to travel for treatment, particularly for palliative radiotherapy. In such instances, after consultation with the patient, radiotherapy may be delivered by a designated practitioner at a radiotherapy centre that is closer to the patient's home. The decision to delegate treatment to a designated practitioner is a balance between the need for radiotherapy, particularly radical, to be planned and delivered in a large volume centre, and the choice of an individual patient to minimize their travelling and inconvenience. Designated practitioners will mostly deliver palliative radiotherapy, and some selected radical radiotherapy.

**Technology**

- **Radiotherapy treatment machines:** Specialised Sarcoma Centres must have

linear accelerators capable of delivering IMRT for patients who need this. Patients best treated with IMRT must not be referred to radiotherapy centres without the capability to deliver IMRT.

- **Proton Beam radiotherapy (PBT):** Specialised Sarcoma Centres must consider PBT for all eligible patients, although it is accepted that some patients may not be able to travel abroad for PBT for personal or practical reasons. When PBT is available in the UK (in 2018), Specialised Sarcoma Centres must consider appropriate referral of all eligible patients for whom PBT is anticipated to offer an advantage over photon radiotherapy, to one of the two designated UK PBT centres.
- **Stereotactic ablative radiotherapy (SABR):** a small minority of patients may be clinically exceptional and suitable for treatment with SBRT. When this is the case, referral must be made to a SBRT treating centre.

**Systemic anticancer therapy services**

Systemic therapies are an important part of the management of sarcomas. Systemic therapies include chemotherapy and other biological agents. The role of chemotherapy may be in a curative or palliative setting.

Chemotherapy is an essential part of management of certain types of bone sarcomas and usually involves complex and intensive therapy regimens. As these intensive treatment regimens require expertise, only designated sarcoma oncologists and paediatric oncologists, who regularly treat these patients, can be responsible for their management.

The role of chemotherapy in soft tissue sarcoma is becoming more complex with the recognition that different histological subtypes may respond differently to various systemic agents.

**Structural**

- **Treatment decisions:** on the use of chemotherapy for individual sarcoma patients is the responsibility of the Sarcoma MDT.
- **Guidance on the use of systemic therapy:** in soft tissue and bone sarcomas is provided in the National Sarcoma Chemotherapy Algorithm.

**People**

- **Chemotherapy** can be given under supervision by clinical, medical and paediatric oncologists who are core members of the Sarcoma or Children's MDT. Most patients will be treated by oncologists within the sarcoma MDT of a Specialised Sarcoma Centre. Radical and palliative treatments will be delivered.
- **Designated practitioners:** are nominated individuals at an oncology centre agreed by their host Trust Clinical Director or equivalent and approved by the SAG to give curative and palliative treatments according to protocols defined by the Sarcoma MDT and SAG. Referrals to designated practitioners must be from an oncologist who is a core member of the Sarcoma MDT.

**Technology**

- The provider of chemotherapy services must provide the facilities for intensive and palliative chemotherapy as described for chemotherapy services in the Manual of

Cancer Services and be either:-

i. A Specialist Sarcoma Centre
ii. A Local Sarcoma Unit
iii. A Children's Cancer Principal Treatment Centre or a Cancer unit for Teenagers and Young Adults
iv. A designated Paediatric Oncology Shared Care Unit.

**Rehabilitation Services**

Patients with sarcoma require rehabilitation. MDTs will have a rehabilitation policy with defined pathways in place and patients must have an individual rehabilitation plan.

**Clinical research infrastructure**

Defining new standards of care through research is an integral part of improving outcomes. The challenges to clinical research include pathological heterogeneity, important biological/genomic differences between sarcoma sub-types, rarity, and the general difficult-to-treat nature of this group of malignancies.

There are clear research opportunities to develop and apply novel trial methodology for smaller trials in individual or groups of histiotypes (rather than continuing the emphasis on large phase III trials randomising all comers). In addition further challenge is set by the dispersal of certain sarcoma subtypes e.g. GIST, to different clinical teams so that patients are not referred to centres where appropriate trials are being run.

All sarcoma MDTs must identify a research lead and SAGs should review research activity regularly. Centres hosting bone MDTs must participate in the activities of the bone sub-group of the NCRI Sarcoma CSG.

All sarcoma patients who may be eligible must be offered the opportunity to take part in clinical trials including tissue donation.

**Pathways for referral and diagnosis:**

SAGs and sarcoma MDTs must work with primary and secondary care providers to agree and implement diagnostic pathways for people with suspected sarcoma.
Through the SAG, diagnostic pathways will be agreed with, and communicated to primary and secondary care providers. These will include details of specialist clinicians for referrers outside the Specialist Centre to liaise with about individual patients (for example, where co-morbidity may preclude travelling to a distant centre or where other clinical complexities exist.

**Suspected soft tissue sarcomas**

All suspected soft tissue sarcomas will be referred to a specialist sarcoma centre, linked diagnostic services at designated local sarcoma units or a Principal Treatment Centre for Children's Cancer according to locally defined and agreed pathways.

Pathways must accommodate tumours arising in all ages and at all anatomic sites.

**Suspected bone sarcomas**

All suspected primary bone tumours will be referred to one of the five designated centres in England.

Diagnosis and surgical resection for bone sarcoma is concentrated in these five centres and each supports a combined bone and soft tissue sarcoma MDT.

The five centres are responsible for the diagnosis of suspected primary bone tumours. These include all malignant tumours and aggressive benign tumours such as giant cell tumours and aneurysmal bone cysts.

SAGs are responsible for ensuring that clear pathways exist for the referral of all suspected bone sarcomas to designated bone tumour services.

Pathways must accommodate tumours arising in the chest wall, spine and craniofacial bones including base of skull. These pathways may recommend surgery for certain anatomical locations to be conducted on sites outside of the five designated centres.

These centres must publish information about their services.

Diagnosis of primary bone tumours made outside the five centres (i.e. inadvertent biopsy or inappropriate treatment) will be the subject of on-going audit between the five centres and the results will be disseminated to Sarcoma Networks.

**Treatment pathways**
**Bone sarcoma**
All adults, children and young people with bone sarcoma must have their care plan confirmed by a bone Sarcoma MDT and treatment delivered by services designated by the SAG.

In cases where chemotherapy is delivered outside of the Specialist Sarcoma Centre, the bone sarcoma MDTs must have clear arrangements in place for liaison with designated practitioners and re-discussion at MDTs during treatment (for example using video-conferencing).

Planned surgery will be undertaken by designated surgical members of a Sarcoma MDT.

The management of bone sarcomas will be in accordance with British Sarcoma Group Guidelines. SACT will be delivered in accordance with the national sarcoma chemotherapy algorithm.

Bone sarcoma MDTs will consider referral of primary tumours deemed inoperable for alternative management including proton beam therapy in accordance with national specifications.

Sarcomas arising in the spine are uncommon and present particular challenges for management influenced by patient, age, co-morbidity, presenting features, location in the spine, histological subtype, stage and response to treatment. Bone sarcoma MDTs must incorporate designated expertise for surgical management of spine sarcomas and all cases must be discussed by the MDT.

The management of craniofacial sarcomas including base of skull tumours will require additional expertise from specialist base of skull and head and neck MDTs. These must

be aligned with a bone sarcoma MDT.

Trusts where additional surgical care may be delivered must be designated and would not be expected to exceed two (allowing for meeting the needs of children) per bone sarcoma MDT. This will ensure simplicity of pathways, access to specialists and concentration of multidisciplinary expertise.

Chordoma arise either in the spine, most commonly at skull base/cervical spine or sacrum. There are fewer than 30 cases per year in the UK. International consensus documents recognise the importance of planned multimodality treatment by specialists. Chondrosarcomas also arise at the skull base and in the spine. All chordomas and chondrosarcomas must be referred to a bone sarcoma MDT and care delivered according to SAG-approved pathways.

Consideration must be given at the time of initial diagnosis to the use of proton beam radiotherapy and integration with surgery. Clear pathways with designated base of skull services must be established to ensure early access to diagnostic and treatment expertise.

All new cases of Ewing sarcoma arising in bone must be referred to the National Ewing MDT which is an exemplar for accessing expertise in order to reach consensus on important areas of clinical uncertainty. Its aim is to discuss all new cases of Ewing sarcoma and to make a consensus recommendation to the treating MDT on surgery and radiotherapy options for primary tumour management.

**Limb and trunk sarcomas**

Approximately 40% of soft tissue sarcomas arise in the limb and trunk. These sarcomas form the majority of cases that will be discussed and managed by a Sarcoma MDT. This sub-population includes considerable heterogeneity varying from small subcutaneous tumours carrying an excellent prognosis when treated by surgery alone to large deep high grade sarcomas which are associated with a 5 year survival rate in the order of 30-40%.

All patients must be diagnosed at designated diagnostic centres in a Local Sarcoma Unit or at the Specialist Sarcoma Centre.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Sarcoma MDTs will publish information about their limb and trunk sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed limb and trunk sarcomas, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for limb and trunk sarcomas within the Sarcoma MDT.

Diagnoses of limb and trunk sarcomas made outside of the local pathway i.e. inadvertent or unplanned biopsies/excision, will be the subject of on-going audit and the results disseminated in the Sarcoma Network.

Adults with soft tissue sarcoma will have their care plan confirmed by a Sarcoma MDT

and treatment delivered by services designated by the SAG. Planned surgery will be undertaken by core or designated members of a Sarcoma MDT.

Management of limb and trunk sarcomas within Specialist Sarcoma Centres will be in accordance with British Sarcoma Group Guidelines.

Atypical lipomatous tumours are common low grade tumours closely related to benign lipomas. Surgery is frequently curative and recurrence, if it occurs, can be successfully managed in most cases by repeat surgery. The risk of metastatic disease is extremely low. Therefore these tumours can be appropriately managed on an 18 week pathway.

**Head and neck sarcoma**

The annual incidence of head and neck sarcoma is approximately 190 soft tissue sarcomas and 38 bone sarcomas, accounting for less than 2% of all head and neck cancers.

There are 49 head and neck MDTs responsible for serving an annual new case load of approximately 9000 laryngeal and oral carcinomas. These range from MDTs seeing less than 30 to others that see more than 100 patients per annum.

Head and neck sarcomas will not be evenly distributed between MDTs. For example, those co-located in Specialist Sarcoma Centres may be more likely to be referred head and neck sarcomas.

Due to the relative rarity of head and neck sarcomas, specialist care cannot be delivered by all head and neck services. SAGs must designate no more than one head and neck service with associated reconstructive support in each Network to manage sarcomas according to pathways determined by the SAG.

Patients with suspected head and neck sarcoma must be referred according to local Sarcoma Network guidelines to a designated diagnostic centre, which will be a Sarcoma Specialist Centre where head and neck and sarcoma MDTs work jointly and patients can access all necessary expertise and support.

People with head and neck sarcomas must have their care plan confirmed by a Sarcoma MDT and treatment delivered by services designated by the SAG.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Sarcoma MDTs will publish information about their head and neck sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed head and neck sarcomas, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for head and neck sarcomas within the Sarcoma MDT.

Diagnoses of head and neck sarcomas made outside of the local pathway i.e. inadvertent or unplanned biopsies/excision, will be the subject of on-going audit and results disseminated in the Sarcoma Network.

Management of head and neck sarcomas within Specialist Sarcoma Centres will be in accordance with British Sarcoma Group Guidelines.

**Gynaecological sarcoma**

Approximately 280 new cases of gynaecological sarcomas are diagnosed annually compared with an annual incidence of approximately 18,000 carcinomas of gynaecological origin.

A gynaecological cancer MDT will on average see about 1 sarcoma for every 100 other cancers. The biology and clinical behaviour of the main types of gynaecological sarcoma all differ markedly from tumours of epithelial origin and expertise in gynaecological sarcoma will not be prevalent in most gynaecological cancer MDTs.

The tumours are mainly uterine but can also rarely arise from the cervix, ovary, fallopian tube, vagina and vulva. The commonest are leiomyosarcomas (low, intermediate and high grade). Other diagnoses include endometrial stromal sarcoma, undifferentiated uterine sarcomas, rhabdomyosarcomas and perivascular epithelioid (mesenchymal) cell tumours (PEComas). Carcinosarcomas are outside this specification following their molecular classification by the Tumour Genome Atlas as having most similarity to high grade epithelial gynaecological tumours. The exception is where sarcoma is the predominant histological component of a tumour.

About two-thirds of gynaecological sarcomas are not diagnosed pre-operatively but discovered on histopathology after hysterectomy which has been carried out for presumed fibroids or other benign conditions. Immediate referral according to the Sarcoma Network pathway to the Sarcoma MDT must be undertaken.

Features on imaging which are atypical for fibroids should raise the suspicion of sarcoma and prompt liaison with the identified sarcoma specialists responsible for gynaecological sarcomas within the Sarcoma MDT.

It is recognised that distinguishing between fibroids and tumour on imaging is very difficult. Laparoscopic morcellation has grown in popularity for the treatment of fibroids because it is associated with lower morbidity than with open surgical procedures. However, morcellation of a uterine sarcoma is associated with higher rates of abdomino-pelvic recurrence, and poorer progression free and overall survival than definitive surgery with hysterectomy. It is therefore contra-indicated for uterine sarcoma.

In many cases the initial management is by total abdominal hysterectomy. This may be undertaken outside the Specialist Sarcoma Centre within a designated gynaecological oncology service after patients have been staged for sarcoma.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Patients with proven gynaecological sarcoma must be referred according to local Sarcoma Network guidelines to a Specialist Sarcoma Centre where gynaecological and sarcoma MDTs work jointly to ensure patients can access all necessary expertise and support.

People with gynaecological sarcomas must have their care plan confirmed by a Sarcoma MDT and treatment delivered by services designated by the SAG.

Sarcoma MDTs will publish information about their gynaecological sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed gynaecological sarcomas, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for gynaecological sarcomas within the Sarcoma MDT.

Management of gynaecological sarcomas within Specialist Sarcoma Centres will be in accordance with national Guidelines.

**Intra-abdominal including retroperitoneal sarcoma**

About 7% of sarcomas are of gastrointestinal origin, a further 6% arise in the abdominal cavity and 5% are retroperitoneal. These sarcomas present particular challenges both to diagnose and treat, often presenting late as large tumours.

Any intra-abdominal or retroperitoneal tumour arising in adults which is not epithelial in origin must be considered to be a sarcoma.

Pre-operative/treatment biopsy is safe and must be considered when the diagnosis is uncertain. Diagnostic biopsy is necessary for tumours that are not demonstrated to be liposarcoma on imaging and often requires specialist trained interventional radiological input.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Surgery remains the primary treatment for these tumours, and is often complex and usually involves multi-visceral/structural resection with careful planning within a multi-specialist surgical team. Best outcomes are achieved with complete surgical resection of these tumours without breach of the tumour capsule or fragmentation of the tumour.

Radical resection often necessitates multi-organ resection and is associated with improved local control/oncological outcomes. However multi-visceral resection carries the potential for increased surgical morbidity and mortality and the best outcomes for these patients are achieved in hospitals that routinely manage complex surgical patients and when surgery is undertaken by surgical teams who specialise in retroperitoneal sarcomas with a high caseload.

Specialist intensive care facilities with 24/7 medical cover, specialist anaesthetic and nursing staff, 24 hour emergency theatre and interventional radiology are required.

The SAG will define sites where surgery for retroperitoneal, abdominal and pelvic sarcomas must be undertaken.

Sarcoma Networks will publish pathways so that people with retroperitoneal sarcoma are referred before any intervention to a sarcoma treatment centre with special expertise in managing this type of tumour. Special expertise will be defined by:-

1) regular sarcoma MDT meetings which include anatomic expertise in pathology,

imaging and surgery
2) infrastructure and resource to support major intra-abdominal surgery
3) a case load including surgical resection of an average of 24 new cases of primary retroperitoneal sarcoma per annum.

All retroperitoneal, abdominal and pelvic sarcomas must be discussed pre-operatively by the sarcoma MDT. Planned surgery will be undertaken by core or designated members of a Sarcoma MDT.

Where surgery is performed inadvertently outside of the specialised sarcoma centre, patients must be referred post-operatively to the specialised sarcoma centre. The SAG must maintain a prospective audit of such cases.

Specialist Sarcoma Centres designated to undertake retroperitoneal sarcoma surgery must have in place a process for discussion with another centre of patients with newly diagnosed retroperitoneal sarcoma deemed inoperable for reasons other than co-morbidity,.

Whilst surgery remains the standard treatment of these tumours, preoperative radiotherapy is increasingly considered for retroperitoneal sarcomas with the development and use of IMRT providing accurate therapy with reduced normal tissue toxicity to adjacent organs. The Sarcoma MDT meetings at which abdominal and retroperitoneal sarcomas are discussed must include appropriate expertise to address questions of radiotherapy and systemic therapy.

Sarcoma MDTs will publish information about their abdominal and retroperitoneal sarcoma pathways, including where these may involve travel to a more distant Specialist Sarcoma Centre and provide liaison points for referring clinicians to discuss suspected or confirmed abdominal and retroperitoneal sarcomas, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for abdominal and retroperitoneal sarcomas within the Sarcoma MDT.

Diagnoses of abdominal and retroperitoneal sarcomas made outside of the local pathway i.e. inadvertent or unplanned biopsies/excision, will be the subject of on-going audit and results disseminated within the Sarcoma Network.

**GIST**
Gastro-intestinal stromal tumours are mesenchymal tumours of the GI tract. They most often occur in patients over 60 but can also uncommonly occur in younger people including children.

There are approximately 700 new cases per year in the UK. Presentation is generally with bleeding or pain with most primary tumours arising in the stomach but also affecting small bowel and other intra-abdominal sites. There is variation in behaviour which is to an extent predicted by size, histological features and tumour DNA mutational analysis.

About 60% of patients will be successfully treated by surgery. Others may have more aggressive sarcomas and benefit from tyrosine kinase inhibitors, principally imatinib, targeting the KIT oncogene and other molecular abnormalities.

GISTs most often present to gastrointestinal services. There is a need to reduce national variation in whether patients with GIST are discussed in sarcoma MDTs or only within GI MDTs. There is also variation in the use of mutational testing which may guide SACT and further variation as to whether patients are referred to specialist sarcoma centres for SACT.

Patients with suspected GIST must be referred according to local Sarcoma Network guidelines to a designated diagnostic centre. People with GIST must have their care plan confirmed by a Sarcoma MDT and treatment delivered by services designated by the SAG. The medical management of patients with GIST must be supervised by cancer specialists with experience in the management of patients with GIST, defined by an annual minimum new case load to a service of 24 per annum, participation in national audit, contribution to national/international clinical trials when available and by being core or designated members of a sarcoma MDT.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and molecular analysis.

Sarcoma MDTs will publish information about their GIST sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed GIST, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for GISTs within the Sarcoma MDT.

**Breast sarcoma**

Soft tissue sarcomas may arise de novo within the breast or as a consequence of previous radiotherapy for breast cancer. The latter are always angiosarcoma. De novo breast sarcomas must be managed as with other soft tissue sarcomas based on stage and histological subtype. Surgery may be appropriately undertaken within breast cancer services after discussion has occurred with a sarcoma MDT. Angiosarcomas must always be referred at suspected diagnosis to a Specialist Sarcoma Centre.

Women with suspected breast sarcoma must be referred according to local Sarcoma Network guidelines to a designated diagnostic centre, which will be a Sarcoma Specialist Centre where breast and sarcoma MDTs work jointly and patients can access all necessary expertise and support. Women with breast sarcomas must have their care plan confirmed by a Sarcoma MDT and treatment delivered by services designated by the SAG.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Sarcoma MDTs will publish information about their breast sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed breast sarcomas, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for breast sarcomas within the Sarcoma MDT.

Phyllodes tumours are rare fibro epithelial tumours arising in the breast which range from benign to malignant. Referral to Specialist Sarcoma Centres is indicated for malignant or

metastatic tumours.

**Skin sarcoma**

Sarcomas of the skin have a more favourable prognosis than either subcutaneous or deep soft tissue sarcomas. However aggressive subtypes such as angiosarcoma occur and other histologies have a tendency for local recurrence leading to additional treatment morbidity if initial management is inadequate.

Patients with suspected skin sarcoma must be referred according to local Sarcoma Network guidelines to a designated diagnostic centre, which will be a Sarcoma Specialist Centre where skin and sarcoma MDTs work jointly and patients can access all necessary expertise and support. People with skin sarcomas must have their care plan confirmed by a Sarcoma MDT and treatment delivered by the Sarcoma MDT or by services designated by the SAG.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Sarcoma MDTs will publish information about their skin sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed skin sarcomas, for instance where co-morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for skin

**Chest wall and lung sarcoma**

Sarcomas may arise either in soft tissue or bone of the chest wall. The principles of management of sarcomas at other sites apply but surgical expertise for complex chest wall resection and reconstruction may not be found within the core surgical members of a sarcoma MDT. SAGs in conjunction with sarcoma MDTs must therefore agree pathways and locations of care appropriate to this tumour location.

All people with suspected or proven bone sarcomas of the chest wall must be referred to a bone sarcoma MDT. Bone sarcoma centres must have clear pathways defining access to thoracic surgery. SAGS must designate no more than one thoracic surgery centre to link to the bone sarcoma MDT.

Primary sarcomas of the lung parenchyma are very uncommon and are commonly diagnosed at the time of resection of an unidentified mass. If the diagnosis is suspected or proven on biopsy, referral to a Specialist Sarcoma Centre is required in accordance with local published pathways.

People with chest wall and lung sarcomas must have their care plan confirmed by a Sarcoma MDT and treatment delivered by services designated by the SAG.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and any appropriate molecular analysis.

Sarcoma MDTs will publish information about their chest wall and lung sarcoma pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or confirmed chest wall or lung sarcomas, for instance where co-

morbidity may preclude travelling to a distant centre, with identified sarcoma specialists responsible for chest wall and lung sarcomas within the Sarcoma MDT.

Metastatectomy and newer approaches such as percutaneous ablation of pulmonary metastases or SABR are important components of managing sarcomas. Sarcoma MDTs must define pathways for patients requiring these approaches which allow multidisciplinary discussion and routine audit of procedures and outcomes.

Metastatectomy and ablation must be undertaken in centres with sufficient caseload volume to ensure at least one case per month in order to ensure maintenance of expertise throughout the MDT. Procedures must be undertaken by core or designated members of the MDT.

**Fibromatosis**

Desmoid Fibromatosis (DF) will often be diagnosed as a result of assessment in a sarcoma diagnostic service. Biopsy is usually necessary to confirm the diagnosis.

At diagnosis or suspected diagnosis, all patients with suspected DF must be referred to the Specialist Sarcoma Centre for diagnosis and management.

Inadvertent resection with positive margins is a frequent feature. In this event, re-resection must not be undertaken and the patient must be referred to the Specialist Sarcoma Centre for diagnosis and management.

Pathology must be referred to a Specialist Sarcoma Pathologist for diagnostic confirmation and appropriate mutational analysis.

In all cases of intra-abdominal fibromatosis, on diagnosis, colonoscopy to rule out Familial Adenomatous Polyposis must be considered. Referral must also be considered in extra-abdominal cases especially if there is a family history of colorectal cancer.

A period of active surveillance is usually appropriate in the first instance. Adjustment to any existing hormone treatment may be necessary prior to commencing active surveillance. Surveillance will usually be in the form of serial MRI scans which must be performed at the sarcoma centre or to the sarcoma protocol at that centre. Subsequent scans must be performed to the same protocol to enable direct comparison.

In the event of disease progression then different treatment options are available. The choice of treatment must be determined by the Sarcoma MDT after discussion with the patient and be in accordance with UK and International guidelines.

Sarcoma MDTs will publish information about their DF pathways and provide liaison points for clinicians outside the Specialist Sarcoma Centre to discuss suspected or DF with identified sarcoma specialists responsible for DF within the Sarcoma MDT.

**Pathways for very rare sarcoma**

**Cardiac/large vessel sarcomas**

Intimal sarcomas and angiosarcomas of the heart and great vessels are a rare entity demanding specific multidisciplinary expertise in order to consider the options for an

individual of combined modality treatment including cardiac surgery.

**Paediatric, wild type and syndromic GIST (PAWS-GIST)**
GISTs very infrequently occur in children and young people. Other GISTs have uncommon molecular profiles or are associated with other clinical manifestations as part of specific syndromes. In these situations, treatments are more frequently ineffective than in other GISTs. Expertise for managing these diseases will not be equally available in all Specialist Sarcoma Centres and referral to centres with special expertise, largely based on a critical mass of patients and multidisciplinary expertise is required.

**Craniofacial bone sarcomas**
Craniofacial bone sarcomas include osteosarcoma, Ewing sarcoma, mesenchymal chondrosarcoma and other less common subtypes.

Management requires close cooperation between radiologist, pathologist, oncologist and surgeons as well as experience in supporting patients though the particular challenges experienced. The care of these very rare tumours must be in centres which have comprehensive teams for bone sarcoma and head and neck cancer surgery and where MDTs have experience based on sufficient case volume of at least ten new cases per year.

**Skull base sarcoma**
The commonest sarcomas arising in the skull base region are chordoma and chondrosarcoma. Some rarer bone sarcomas such as Ewing sarcoma and soft tissue sarcomas can involve the skull base. There number of cases in England each year is currently hard to determine with great accuracy but likely to be less than 40 per year. There are key inter-dependencies which include disease familiarity and procedure expertise for both surgical and oncological team members and an increasingly essential link to proton beam therapy (PBT) services. Therefore management of these rare sarcomas must be focussed at a very small number of centres, likely no more than five in England, where very close interaction between skull base unit and sarcoma MDT can be ensured with detailed examination of outcomes.

**Spinal, Paraspinal, Complex Pelvic and Sacral Sarcoma involving bone**
These are a heterogeneous group of rare sarcomas that require highly specialised multidisciplinary assessment, treatment planning and delivery. Where possible, primary surgery remains the gold standard but is often highly morbid and recurrence common. Newer highly specialised techniques such as computer-assisted resection may improve the safety of resection. PBT has increasing importance, either as a pre- or post-operative adjunct to surgery or in some cases instead of surgery. This can require specific consideration of stabilisation techniques and post-operative PBT treatment planning.
Suspected sarcomas at these sites must be referred to a designated bone sarcoma service. Surgery must be undertaken by core members of a bone sarcoma MDT.

**Children with sarcoma**
All children with suspected bone sarcomas must be referred to a bone tumour diagnostic service.

Up to the age of 10 years, the commonest soft tissue sarcoma in children are rhabdomyosarcomas which may arise at several anatomical sites. Other histological

subtypes of soft tissue sarcoma are much less frequent until about 10 years when 'adult-type' histologies are of similar incidence to rhabdomyosarcoma. Although paediatric oncology MDTs will have core expertise for management of most rhabdomyosarcomas, early discussion within a month of diagnosis with sarcoma MDTs is required for extremity tumours and all non-rhabdomyosarcomas for whom the benefits of specialist diagnostics and surgery may be considerable.

Surgeons operating on children with sarcomas must be core or designated members of a sarcoma MDT. SAGs must ensure that Children's Cancer MDTs and Sarcoma MDTs have systems in place to ensure effective communication.

**Links to TYA MDTs**

All teenagers and young adults' referrals must be discussed at the appropriate TYA MDT for decision on care planning and treatment with clear mechanisms for tracking. The Bone Tumour MDT must be integrated with a Teenage & Young Adult (TYA) Principle Treatment centre that meets the Improving Outcomes Guidelines for Teenagers & Young Adults (2001). The operational policy must have the working arrangements between these MDTs clearly stated and agreed with host networks and NHS England commissioners.

**Follow up pathways**

For most high grade sarcomas, recurrence risk is highest in the first 2-3 years after completion of treatment. For others, recurrence is well-recognised to occur even after many years of disease-free survival and follow-up schedules and treatments, must be guided by sarcoma sub-type. Patients must be educated at the end of treatment about the potential manifestations of recurrence and advised about self-detection of local recurrence. Routes to re-access clinical teams must be clear and supported by written information in an end of treatment summary.

All SAGs must agree in conjunction with Sarcoma MDTs follow-up guidelines and also define where follow up is appropriately undertaken in Local Sarcoma Units.

For some patients who are successfully treated, the morbidity including long term sequelae is considerable. The survivorship needs of sarcoma patients must be addressed by appropriate surveillance schedules and support.

**Rehabilitation pathways**

Sarcoma surgery has short and long term impact on patients in different ways depending on the site and complexity of the surgery undertaken. For these reasons good access to rehabilitation services and supportive care are particularly relevant to sarcoma patients.

It is therefore important that patients with sarcoma are supported from diagnosis through the entire pathway by access to appropriate rehabilitation resources.

The rehabilitation care pathways provide a model for this support and cover the acute, community and primary care settings. There should be appropriate assessment of patients' rehabilitative needs across the pathway and the provider must ensure that high quality rehabilitation is provided in line with the network agreed sarcoma rehabilitation pathway.

A specialist sarcoma physiotherapist and other specialised AHPs will be members of the

sarcoma multidisciplinary team

Ongoing rehabilitation and supportive care will be provided locally wherever possible. This will be co-ordinated by the therapist in liaison with the key worker.

Patients with functional disabilities as a consequence of their sarcoma must have timely access to appropriate support, rehabilitation and limb fitting services if appropriate.

**Symptom care and end of life care pathways**
People with sarcoma can present a range of challenges for effective symptom relief and meeting of complex end of life needs. Sarcoma MDTs must have effective communication and referral processes with associated hospital and community palliative care services.

### 3.3 Population covered

This specification is for all patients with suspected or diagnosed bone and soft tissue sarcoma. This includes fibromatosis and other soft tissue tumours of borderline or intermediate malignancy (e.g. solitary fibrous tumours, hemangioendotheliomas and similar vascular tumours, PEComas).

The specification also includes the surgical and other treatment of giant cell tumour of bone. The surgical treatment of primary lymphoma of bone and solitary plasmacytoma are also included.

The specification covers the care of patients with gastrointestinal stromal tumours and desmoid fibromatosis at any site.

### 3.4 Any acceptance and exclusion criteria and thresholds

The service outlined in this specification is for patients registered with a GP practice in England.

Soft tissue sarcoma MDTs, unless they are a joint soft tissue/bone sarcoma MDT, are not commissioned to provide the investigation and the surgical treatment of malignant primary tumours of the bone.

The specification does not cover treatment for confirmed benign primary bone tumours other than giant cell tumour of bone, haematological conditions such as multiple myeloma, lymphoma involving bone.

Carcinosarcomas are outside this specification following their molecular classification by the Tumour Genome Atlas as having most similarity to high grade epithelial gynaecological tumours. The exception is where sarcoma is the predominant histological subtype.

Kaposi's sarcoma is not covered by this specification given its likely endothelial origin and

limited overlap with sarcoma care.

### 3.5 Interdependencies with other services/providers

Sarcoma services are commissioned as part of a network arrangement that must include a specialist sarcoma centre hosting an MDT.

A comprehensive service is available for patients with any type of sarcoma and network pathways are the responsibility of the network's SAG.

The network pathways are developed with the sarcoma MDT, Local Sarcoma Units and other centres which are designated within the network for the delivery of diagnostic and treatment services.

The sarcoma MDT is hosted at a specialised sarcoma centre and is responsible for the management of soft tissue sarcoma of a combined MDT for soft tissue and bone sarcoma.

The SAG is the primary source of clinical opinion for sarcoma services and is also responsible for clinician and local sarcoma unit designation.

The specialist sarcoma unit is designated by the SAG to host the Sarcoma MDT. The local sarcoma unit is designated by the SAG to deliver defined elements of care for sarcoma patients.

The designated practitioner is an extended member of the sarcoma MDT delivering specific elements of sarcoma care.

Members of the specialist sarcoma MDT, the local sarcoma unit and designated practitioners must contribute to and support the work programme of the SAG.

The SAG and the sarcoma network will engage with local health, social care and other services to ensure that patient experiences across all aspects of sarcoma pathways (specialised and non-specialised) are positive.

For intra-abdominal including retroperitoneal sarcoma and other major sarcoma surgical procedures, specialist intensive care facilities with 24/7 medical cover, specialist anaesthetic and nursing staff, 24 hour emergency theatre and interventional radiology are required.

## 4. Applicable Service Standards

### 4.1 Applicable national standards e.g. NICE

Improving Outcomes for People with Sarcoma NICE (2006)

Improving Outcomes for Children and Young People NICE (2005)

Manual for Cancer Services Sarcoma Measures Department of Health (August 2011)

Manual for Cancer Services Acute Oncology Measures Department of Health. (April 2011)

Manual for Cancer services Chemotherapy Measures (April 2013)

NICE Quality Standard for Sarcoma

### 4.2 Applicable standards set out in Guidance and/or issued by a competent body (e.g. Royal Colleges)

British Sarcoma Group Guidance for Soft Tissue Sarcoma

British Sarcoma Group Guidance for Bone Sarcoma

UK Guideline for Gastrointestinal Stromal Tumour (GIST)

## 6. Location of Provider Premises

**Joint Soft Tissue and Bone Sarcoma MDT Providers**

The Newcastle Upon Tyne Hospitals NHS Foundation Trust

The Royal Orthopaedic Hospital NHS Foundation Trust

London Royal National Orthopaedic Hospital NHS Trust with University College London Hospitals NHS Foundation Trust

Oxford University Hospitals NHS Trust

Robert Jones and Agnes Hunt Orthopaedic and District Hospital NHS Trust

**Soft Tissue MDT Providers**

University Hospital Birmingham NHS Foundation Trust

North Bristol NHS Trust and University Hospitals Bristol NHS Foundation Trust

Plymouth Hospitals NHS Trust

Royal Devon and Exeter NHS Foundation Trust

The Christie NHS Foundation Trust and Central Manchester University Hospitals NHS Foundation Trust

Royal Liverpool and Broadgreen University Hospitals NHS Trust Compliant

Nottingham University Hospitals NHS Trust and University Hospitals Of Leicester NHS Trust

The Royal Marsden NHS Foundation Trust

Leeds Teaching Hospitals NHS Trust

Sheffield Teaching Hospitals NHS Foundation Trust

**Appendix 1**

**Guidance, measures and standards**

Improving Outcomes for People with Sarcoma (NICE 2006)
https://www.nice.org.uk/guidance/csg9

Manual for Cancer Services, Sarcoma Measures version 1, January 2014
https://www.cquins.nhs.uk/./measures/Sarcoma_January2014.pdf

Improving Outcomes in Children and Young People with Cancer (NICE August 2015)
https://www.nice.org.uk/Guidance/csgcyp

NICE Quality Standard for Sarcoma (NICE January 2015)
https://www.nice.org.uk/guidance/qs78

Cancer Waiting Times
http://www.nhs.uk/choiceintheNHS/Rightsandpledges/Waitingtimes/Pages/Guide%20to%20waiting%20times.aspx

NICE Suspected cancer: recognition and referral
http://www.nice.org.uk/guidance/ng12

**Appendix 2**

**Sarcoma Metrics linked to the aim and objectives**

| **Sarcoma metrics linked to the aim and objectives** | |
|---|---|
| All patients with a newly diagnosed sarcoma will be referred to a specialist sarcoma service.<br>To provide a comprehensive service for all patients with sarcoma<br>To ensure compliance with Care Quality Commission regulations | |
| **Service specification objective** | **Indicator** |
| To manage the care of at least 100 new patients with soft tissue sarcoma per year (soft tissue sarcoma MDT)<br>To manage the care of at least 50 new patients with bone sarcoma per year and least 100 new patients with soft tissue sarcoma per year (joint soft tissue and bone sarcoma MD | The number of new cases per year |
| To provide expert accurate diagnosis and staging of bone and soft tissue sarcoma utilising the most up to date validated diagnostic tools<br><br>To have pathways for referral and diagnosis in place for people with suspected sarcoma<br><br>To publish information about shared pathways, activity and patient outcomes, including information on site specific sarcomas | % Patients without confirmed histology prior to surgery<br><br>% of biopsies outside designated centre for soft tissue sarcoma<br><br>% of biopsies outside designated centre for bone sarcoma<br><br>Proportion of patients whose sarcoma is staged using the TNM staging system prior to definitive treatment<br><br>% patients with time to diagnosis within 30 days<br><br>% patients meeting RT waiting times<br><br>No. of patients with a second malignancy<br><br>There are network agreed patient pathways in place<br><br>Proportion of children with non-rhabdomyosarcoma histologies and limb/trunk rhabdomyosarcomas |

| | |
|---|---|
| | discussed within 1 month of diagnosis at sarcoma MDT<br><br>% of TYA patients discussed at TYA MDT |
| To ensure that all patients with confirmed bone of soft tissue sarcomas have access to expert care and management provided by trained staff and receive care in accordance with the most up to date agreed clinical protocols<br><br>To provide chemotherapy, radiotherapy and surgery in line with national guidelines, evidence based practice and network policy and delivered by designated practitioners who are core or extended members of a Sarcoma MDT.<br><br>To be compliant with National Guidance and standards including NICE IOG and standards<br><br>To support local sarcoma units to manage patients with sarcomas in line with national guidelines, evidence based practice and network policy | There are network wide clinical guidelines in place which, where available, reflect national guidelines and policy<br><br>There are designated practitioners who are named members of the Sarcoma MDT delivering elements of care in Local Sarcoma Units outside the specialist sarcoma centres<br><br>% of patient receiving radiotherapy at designated local treatment centres<br><br>% of patient receiving chemotherapy at designated local treatment centres<br><br>Amputation rates for extremity bone and soft sarcoma<br><br>Proportion of patients with extremity sarcoma, who undergo curative surgical resection where R0 or planned R1 resection is achieved<br><br>Proportion of patients with an extremity soft tissue sarcoma which is deep and grade 2 or 3 who receive post-operative radiotherapy within 3 months of a planned marginal or wide local excision (R0 or R1).<br><br>% patients treated with RT pre-operatively extremity soft tissue<br><br>% patients treated with IMRT<br><br>No. patients receiving chemotherapy outside national algorithm<br><br>Proportion of patients with high or moderate risk GIST, small bowel GISTs and primary metastatic GIST who have mutational analysis within 3 months of diagnosis<br><br>Proportion of patients with high/moderate |

| | |
|---|---|
| | risk GIST who commence adjuvant imatinib within 3 months of complete macroscopic resection |
| For adults, children and young people with bone sarcoma and adults with soft tissue sarcoma – to have their care plan confirmed by a sarcoma MDT and treatment delivered by services designated by the Sarcoma Advisory Group | There is an MDT that meets the requirements as specified in the national service specification<br><br>There is a weekly MDT meeting for treatment planning attended by all the relevant disciplines<br><br>% patients discussed at Sarcoma MDT prior to definitive treatment<br><br>% patients discussed at an MDT prior to definitive treatment<br><br>% excisions outside designated centre<br>Number of patients referred for PBRT |
| To ensure optimal physical functioning and quality of life for patients through rehabilitation and monitoring | |
| To ensure there is a commitment to continued service improvement and to improve outcomes for patients with sarcoma through research and audit | Survival - 1 and 5 year<br><br>Local recurrence rate for extremity soft tissue sarcomas<br><br>Local recurrence rate for extremity bone sarcomas<br><br>No. patients who died within 30 days of treatment<br><br>% of patients accepted entry into a clinical trial |
| To support patients through an allocated key worker with specialist knowledge of sarcomas and their treatments<br><br>To provide high quality information for patients, families and carers in appropriate and accessible formats and mediums | % patients seen by a Sarcoma CNS<br><br>Proportion with recorded treatment intent |

| | |
|---|---|
| To ensure that there is involvement of service users and carers in service development and review | |
| To present information to the annual audit meeting (joint soft tissue and bone sarcoma MDT) | |

**Appendix 3**

Designated MDT members and designated practitioners

A sarcoma MDT includes core members who fulfil the requirements of expertise, allocated time in job plan and attendance. Comprehensive sarcoma care is dependent on the support to the core MDT provided by other practitioners who by way of expertise or location can positively contribute to high quality specialist care. Examples include (i) surgeons whose main anatomical focus is not sarcoma but may for individual cases be the most appropriate lead or support surgeon, and (ii) oncologists able to deliver radiotherapy or SACT under the direction of the sarcoma MDT nearer to the patient's home. These latter roles are described as 'designated practitioners' in the Manual of Cancer Services for Sarcoma which also describes qualifying criteria to be fulfilled. This includes being a member of an extended sarcoma MDT. Although the term 'extended MDT member' is widely used it is loosely defined.

In this specification, all practitioners who may be involved in delivery of care for patients with sarcoma must be designated to do so by the SAG in conjunction with the sarcoma MDT. The principle of designated practitioner will therefore be extended beyond oncologists to include surgeons and other specialists. This is also essential for the monitoring the delivery of high quality specialist care.

Designation as a member of the extended sarcoma MDT will require

- Nomination by the sarcoma MDT
- A written record shared between sarcoma MDT, SAG and employing Trust clinical line manager (clinical or Medical Director)
- An outline of role as a designated practitioner e.g.
    - to deliver chemotherapy in accordance with Network guidelines and following recommendation for individual patient from the sarcoma MDT
    - to be responsible for surgical management of (site specific) sarcoma in accordance with Network guidelines and following recommendation for an individual patient from the sarcoma MDT
- Designated oncologists will be expected to participate directly in sarcoma MDT meetings either on a regular basis or for case-by-case discussion.
- Participation in Sarcoma MDT annual review
- Participation in pathway development and maintenance
- Participation in audit of sarcoma patients
- Named in information made available to patients by sarcoma MDT and SAG
- Designation reviewed by Sarcoma MDT and SAG at least biennially.