

B12/S(HSS)/a

**2013/14 NHS STANDARD CONTRACT  
FOR PRIMARY MALIGNANT BONE TUMOURS SERVICE (ADULTS AND  
ADOLESCENTS)**

**PARTICULARS, SCHEDULE 2 – THE SERVICES, A – SERVICE SPECIFICATION**

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| <b>Service Specification No.</b> | B12/S(HSS)/a  |
| <b>Service</b>                   | Primary malignant bone tumours service (adults and adolescents) |
| <b>Commissioner Lead</b>         |   |
| <b>Provider Lead</b>             |   |
| <b>Period</b>                    | 12 months   |
| <b>Date of Review</b>            |   |

## 1. Population Needs

### 1.1 National/local context and evidence base

Malignant primary bone tumours are rare with an overall incidence around 10 new cases per year per million total population. The most common tumours are:

- Osteosarcoma (3 per million)
- Chondrosarcoma (2 per million)
- Ewing's sarcoma (1.5 per million)

The remainder are a variety of other sarcomas. Multiple myeloma, although arising from the bone marrow, is not regarded as a primary bone tumour as its treatment is mainly haematological.

Most patients with bone tumours present with pain (particularly at night or non-mechanical) and/or swelling. Pathological fracture is a less common mode of presentation. It is not uncommon for symptoms to have been present for some time before medical attention is sought or further investigations/referrals are initiated. A full radiological and histopathological/pathological classification is essential to plan treatment. A biopsy is nearly always required and should be assessed by an expert pathologist. Following staging, the multidisciplinary team (MDT) should discuss the case for radiological and pathological correlation and agree the final diagnosis and stage of disease. Treatment can be subsequently planned, agreed and carried out.

The treatment required is dependent on diagnosis and stage of the disease. Treatment modalities used include surgical treatment and/or oncological treatment

such as chemotherapy and radiotherapy. Surgical treatment is usually very challenging and 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, and an amputation may be required in up to 10% of cases. Following treatment oncological and surgical follow up is required.

The prognosis is dependent on the diagnosis, stage of the disease and response to chemotherapy. It is usually expressed as five year survival and has been quoted to be between 50 and 90% for patients with an Enneking stage 2B osteosarcoma.

The investigation of suspected primary bone tumours and the surgical treatment of diagnosed malignant primary tumours of bone are funded by NHS England. However, the oncological treatment is not.

### **Evidence base**

This specification draws its evidence and rationale from a range of documents and reviews as listed below:

- Cancer Commissioning Guidance. Department of Health (2008)
- Improving Outcomes (IOG) in Sarcoma Cancers. Department of Health (2006)
- Manual for Cancer Services Sarcoma Measures Department of Health (August 2011)
- Manual for Cancer Services Acute Oncology Measures Department of Health (April 2011)

It is well recognised that the evaluation of musculoskeletal tumours is complex. Multiple radiological investigations are required to come to a provisional diagnosis and stage [1]. Following appropriate staging, the biopsy should be planned and performed in the centre ultimately responsible for the treatment of the patient [2]. Careful planning of the biopsy site increases the diagnostic accuracy. However, more importantly, the biopsy site should be in line with the excision required for surgical treatment so that the biopsy track can be excised at the time of definitive surgery. The biopsy sample should be discussed at MDT meetings which consist of:

- radiologists
- pathologists
- orthopaedic surgeons
- oncological surgeons
- oncologists with the appropriate experience and expertise to plan treatment [3]

The surgical treatment and subsequent reconstruction is complex and requires careful planning and expertise [4-7]. Limb salvage surgery is feasible in around 90% of the cases. Amputation will be required in up to 10% of patients. It is generally accepted that limb salvage does not jeopardise survival and that in the majority of

cases an acceptable function can be obtained. Overall survival is dependent on several factors including diagnosis, grade, stage and response to chemotherapy. It has been reported to be between 60 and 90%.

Surgical reconstruction can be with an endoprosthetic replacement or a biological construct [5-7]. Such as vascularised free fibula graft. In children, the endoprosthesis or construct will also need to accommodate for future growth [8]. With modern surgical treatment, an overall local recurrence rate of around 5-10% can be expected.

The functional outcome is usually measured with the musculoskeletal tumour society (MSTS) score which is an objective score and/or the Toronto extremity salvage score which is subjective [10-12].

Following surgical treatment, oncological and surgical surveillance is required. The patient is followed up at three monthly intervals for the first two years, followed by six monthly intervals up to 10 years, followed by yearly review [13]. Initial follow up is to monitor for local recurrence and/or metastatic disease. Follow up after five years is particularly to monitor the surgical reconstruction for mechanical problems and the late effects of chemotherapy.

## **2. Scope**

### **2.1 Aims and objectives of service**

The overall aim of the service is to improve outcomes and provide the highest quality of care to people with malignant primary tumours of bone (PMBT). The PMBT specialist MDT service will provide assessment, diagnosis and treatment in line with *Improving Outcome Guidance for People with Sarcoma (2006)*, *Improving Outcome Guidance for Teenage & Young Adults (2011)* and *Cancer Waiting Times*. The nationally commissioned service is limited to the surgical management and excludes oncology (and radiotherapy). The nationally designated PMBT centre(s) will manage the care of at least 50 new patients of bone sarcoma per year.

Specifically the PMBT service aims to provide:

- high quality holistic care delivered through a multidisciplinary team including: orthopaedic surgery, radiology, pathology, medical oncology, clinical oncology, physiotherapy and nursing care
- radiological and pathological facilities to classify and stage the condition prior to planning treatment
- to advise and undertake investigations to proceed to surgical treatment options if clinically indicated
- high quality surgical treatment of patients with PMBTs
- long term surveillance after definitive treatment
- continuous monitoring of risk and governance to ensure that clinical treatment is safe and effective

- clinical and service audits to ensure highest standards of safety, care and
- clinical effectiveness against local and national guidelines

The nationally designated centres are required to agree the following areas with NHS England:

- service configuration and population coverage
- develop and agree referral criteria, clinical protocols, network policies and treatment pathways
- actively engage and participate with the network groups for bone sarcoma tumours
- actively engage and participate in peer review for bone sarcoma tumours

The designated centre will be expected to play a key role in developing and supporting the work programme for “sarcoma advisory group” or designated group covering the host cancer networks within their catchment area including Teenager & Young Adult coordination group.

**Objectives:**

- to provide an exemplary and comprehensive service for all eligible referred patients with PMBT that is delivered in line with the improving outcomes guidance and cancer waiting times
- to provide expert diagnosis of PMBT utilising the most up-to-date validated diagnostic tools and knowledge and improve accuracy of diagnosis
- to provide expert care and management of all patients with confirmed PMBT through the use of the most up-to-date clinical protocols and surgical management
- to consider chemotherapy/radiotherapy and where clinically appropriate provision of surgery in line with national guidelines, evidence based practice and treatment pathways
- effective monitoring of patients to ensure optimal functioning and quality of life for the patient with regards to their PMBT
- to operate a rolling programme of clinical audit to test current practice and inform the evolution of care in PMBT
- to provide care with a patient and family centred focus to maximise the patient experience of care within the nationally designated providers
- to be seen as the leading clinical services and a source of expert advice for the diagnosis and management of PMBT within the NHS
- to support local healthcare providers to manage patients with PMBT whenever it is safe to do so and clinically appropriate
- provide high quality information for patients, families and carers in appropriate and accessible formats and mediums
- to develop the experience, knowledge and skills of the MDT to ensure high quality sustainable provision
- to ensure that there is involvement of service users and carers in service development and review

- to ensure there is a commitment to continual service improvement
- to be compliant with peer review cancer measures
- to ensure compliance with Care Quality Commission regulations

## 2.2 Service description/care pathway

The service aims to deliver high quality clinical care to patients with suspected malignant primary tumours of bone. In addition it funds the surgical treatment of patients with diagnosed malignant primary bone tumours.

Radiological and pathological staging and discussion at MDT is essential to plan subsequent treatment. A biopsy is usually required and should be performed by the national designated provider.

Patients can be referred from primary care, emergency departments and secondary care. To avoid delay in making the diagnosis, appropriate radiological investigations are usually performed by the designated provider. Following biopsy, the case is discussed at the MDT.

The national providers core MDT includes:

- orthopaedic and oncological surgeons
- radiologists
- pathologists
- medical oncologists
- clinical oncologists
- paediatric oncologist
- nurses.

In addition to this the service calls on the work of:

- specialist sarcoma physiotherapist
- specialised allied health professionals (AHP)
- paediatric oncologist
- specialist nurse(s)
- affiliated medical or clinical oncologist from linked cancer centre
- affiliated diagnostic service clinicians
- other professionals including orthopaedic, thoracic, plastic, head and neck, gynaecological, gastrointestinal (GI) and vascular surgeon.

All children, 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 PMBT 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.

The designated centres will deliver the service in line with the following:

- the surgical team liaises with the oncologists to plan treatment as required per designated treatment protocols. Individuals work together with the same aims and clinical understanding of the condition and its management to create a multidisciplinary team approach
- inpatients are reviewed daily on a ward round supported by a consultant orthopaedic and oncological surgeon with input from the core MDT as clinically required. Care plans are clearly documented in the notes. Relevant investigations will be carried out. Any referred patients that are waiting for admission are discussed and the plan to admit them as soon as possible is reviewed with any actions required updated
- the timing of surgery is discussed between oncologist and surgeon as appropriate to the designated treatment protocols
- a weekly MDT is led by the orthopaedic and oncological surgeons to discuss the needs of each newly referred patient (and other patients as required) in detail and review other non-surgical aspects of their care
- the providers will hold other meetings regularly through the month to address clinical, service delivery and governance issues
- there are clinical protocols for the oncological treatment of patients with PMBTs
- audit is an integral part of improving the delivery of care and an ongoing audit programme provides the evidence to improve and enhance the delivery of the clinical care we provide

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

The provider of chemotherapy and radiotherapy services should:

- provide the facilities for intensive inpatient chemotherapy and radiotherapy as described in the 'manual for cancer services'
- be either
  - at a sarcoma treatment centre or
  - at a centre with a nominated medical and/or clinical oncologist who is a member of an extended sarcoma MDT and who agrees to give curative and palliative treatments (chemotherapy or radiotherapy) according to protocols defined by the sarcoma MDT. These oncologists should be nominated by the host cancer network clinical director and approved by the lead clinician on the sarcoma MDT or
  - at a principal treatment centre for children or young people as described in the NICE guidance on 'Improving outcomes in children and young people with cancer'

- offer all patients with soft tissue sarcomas entry into the relevant clinical trials
- the sarcoma MDT should recommend the treatment regimen

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

A specialist sarcoma physiotherapist (SSP) and other specialised allied healthcare professionals (AHPs) will be members of the extended sarcoma MDT ongoing rehabilitation and supportive care will be provided locally wherever possible. This will be coordinated by the therapist in liaison with the key worker

patients with functional disabilities as a consequence of their sarcoma should have timely access to appropriate support and rehabilitation services

### **Pathology**

All bone sarcomas will either be first reported or reviewed by an SSP. An SSP is a pathologist who regularly reports bone sarcoma tumours and these form a significant component of their workload. The SSP will participate in the bone tissue pathology external quality assessment (EQA) scheme and be part of a properly constituted sarcoma MDT.

All gastrointestinal stromal tumours (GISTs) will be reported or reviewed by an SSP with experience in GIST who successfully participates in the bone tissue pathology EQA scheme, or a tertiary GI specialist who successfully participates in the gastrointestinal pathology EQA scheme.

All patients with bone sarcoma tumours assessed in a diagnostic clinic will have their pathology reported by: either an SSP or a pathologist nominated by the sarcoma MDT as part of the local diagnostic referral pathway who has formal links to an SSP

All malignant bone sarcoma tumours will be reviewed by an SSP prior to management recommendations by the sarcoma MDT

There will be at least conditional Clinical Pathology Accreditation approval for the laboratory in which the SSP and those with a specialist interest work

there will be formal documented audit of the work of the SSPs and the nominated pathologists

the SSPs will have ready access to molecular pathology and/or cytogenetics facilities

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

### **Palliative Care**

Patients who require palliative care will be referred to a palliative care team in the

hospital and the team will be involved early to liaise directly with the community services.

Patients who are managed by a sarcoma MDT will be allocated a key worker. They will be provided with their key worker's name and contact details.

Specialist palliative care advice will be available on a 24 hour, seven days a week basis.

### **Support and Follow-up**

Access to cancer genetic services should be offered to the patient and their family, where appropriate

The chest x-rays and clinical examinations will be provided at regular intervals

Long-term follow up will be expected for those patients who have received a prosthetic replacement

There will be regular imaging of patients at high risk of recurrence

### **Risk management**

Care delivered by the PMBT service providers must be of a nature and quality to meet the care standards, specification and agreement for the service. It is the trust's responsibility to notify the commissioner on an exceptional basis should there be any breaches of the care standards. Where there are breaches any consequences will be deemed as being the trust's responsibility.

### **Days/Hours of operation**

24 hours a day, 365 days a year

### **Discharge Planning:**

Criteria for discharge from inpatient care:

- no further investigation required
- no adverse outcomes anticipated
- patient is safe post-surgical excision and reconstruction or amputation
- clinically appropriate arrangements for local care and PMBT service follow-up have been discussed and agreed by all relevant parties
- parents/carers have demonstrated competence in any care they will be required to provide in relation to PMBT
- parents/carers understand and have the necessary information to contact their nationally designated PMBT provider

All discharge planning will be managed by the orthopaedic and oncological surgeons in charge of the case with local health and social care providers being fully informed of the patient's condition and any responsibilities they will have to assume. This will

be formalised in writing to the patient's general practitioner and all other relevant parties.

## 2.3 Population covered

This service covers patients registered with an English General Practitioner, resident in the European Union and eligible for treatment in the NHS under reciprocal arrangements. Patients from Scotland, Wales and Northern Ireland are not part of this commissioned service and the trust must have separate arrangements in place.

## 2.4 Any acceptance and exclusion criteria

### Referral criteria, sources and routes

Referrals are accepted from any qualified doctor where the patient has confirmed or suspected PMBT. The receiving clinician at one of the providers may request the referrer to carry out further investigations to aid the proper diagnosis of the patient's condition. Referrals will be accepted by the nationally designated providers via the on-call surgeon for the PMBT service.

**Inclusion criteria:** Including the following tumours:

**osteosarcoma** is a primary sarcoma of bone where the tumour cells directly produce bone. A variety of subtypes of osteosarcoma have been described depending on histological appearance and / or site in the bone  
Medullary osteosarcoma can be high grade or low grade. High grade medullary osteosarcoma is further sub-classified into osteoblastic, chondroblastic, fibroblastic, telangiectatic, small cell and giant cell or epithelioid depending on histological appearance. In addition, osteosarcoma can be secondary to other pathology such as Paget's disease and previous radiotherapy. Li-Fraumeni and hereditary retinoblastoma syndromes are associated with osteosarcoma Surface osteosarcoma is less common than medullary osteosarcoma. Surface osteosarcoma is sub-classified into parosteal, periosteal and high grade surface osteosarcoma

**chondrosarcoma** is a malignant cartilage tumour occurring within the bone. Enchondroma and atypical enchondroma are its benign counterpart. Predisposing factors for malignancy include Ollier's disease (multiple enchondromas), Maffucci's disease and diaphyseal aclasia (multiple osteochondromatosis)  
Dedifferentiated chondrosarcomas are usually low grade or benign tumours that have transformed into a higher grade tumour of a different cell line, i.e. osteosarcoma or high grade pleomorphic sarcoma. Chondrosarcomas are best regarded as a spectrum of disease as their distinguishing features are a continuum. The hallmark that differentiates benign from malignant tumours is the presence of a permeative pattern, whereby host bone is invaded by cartilage tumour. This can be very difficult to demonstrate on a small biopsy of low grade tumours as they can be prone to sampling error (particularly if they have been taken without image guidance).

The national providers are funded for the diagnosis of all cartilage tumours of bone and the surgical treatment of chondrosarcomas, but not for the surgical treatment of enchondromas or atypical enchondroma.

**Ewing's sarcoma** is a small round cell sarcoma that mainly affects children and young adults. The tumour is most common in the pelvis or diaphysis of the femur, tibia and humerus; although any bone can be affected

**giant cell tumour** of bone is an aggressive benign bone tumour with multiple reactive giant cells. Although a benign tumour, its behaviour is aggressive and treatment complex. Therefore, the national providers are also designated for the surgical treatment of this tumour

**chordoma** is a rare malignant primary bone tumour showing notochordal differentiation. It is confined to the spine and the base of skull, and the sacrococcygeal region is the most commonly involved. Chordoma is a disease of adult age, although rare cases have been described in children

**osteofibrous dysplasia and adamantinoma** represent a spectrum of primary bone tumour showing epithelial differentiation. The most extreme end of the spectrum is represented by de-differentiated adamantinoma, in which osteofibrous dysplasia/well differentiated adamantinoma is associated with a high grade sarcoma, which can be an osteosarcoma or an undifferentiated spindle cell sarcoma

**miscellaneous primary bone sarcomas.** This group is represented by other types of rare sarcomas which may primarily arise in bone, including malignant vascular tumour (epithelioid haemangioendothelioma, angiosarcoma, Kaposi sarcoma), and other types of spindle cell sarcomas (myofibrosarcoma, leiomyosarcoma, rhabdomyosarcoma, undifferentiated spindle cell sarcoma)

### **Exclusion criteria**

The national designated PMBT service is not commissioned to provide treatment for:

- confirmed benign primary bone tumours other than giant cell tumour of bone
- multiple myeloma (but solitary plasmacytoma is included)
- lymphoma not primary arising in bone

### **Response time & detail and prioritisation**

Initial telephone contacts from referrers are to be dealt with immediately by the senior PMBT surgeon on duty. The referral may be accepted over the phone immediately and/or the PMBT service provider may request the referrer to carry out further investigations. Advice on optimal management will be given and on-going support will be provided until the patient is transferred.

Transfer of patients to a nationally designated PMBT service will be prioritised according to the needs of individual patients but in all cases where a transfer to the PMBT service has been agreed that transfer will take place as soon as is

practicable. If necessary the nationally designated PMBT providers will communicate with each other to coordinate appropriate care for patients at times when capacity at one of the provider is under pressure.

## **2.5 Interdependencies with other services**

The PMBT team will link into multiple clinical and administrative teams as a result of the composition of the broad MDT.

The nationally designated PMBT providers are the leaders in the NHS for patient care in this area. They provide a direct source of advice and support when other clinicians refer patients into the nationally designated providers. This support will continue until the patient is transferred into the nationally designated provider or it becomes apparent that the patient does not have a PMBT.

The nationally designated providers will also provide education within the NHS to raise and maintain awareness of PMBT and its management.

The national providers will form a relationship with local health and social care providers to help optimise any care for PMBT provided locally for the patient. This may include liaison with consultants, general practitioners, community nurses or social workers etc.

## **Relevant networks and screening programmes**

The national providers form part of the network site specific group for sarcoma within their cancer network.

## **3. Applicable Service Standards**

### **3.1 Applicable national standards e.g. NICE, Royal College**

The nationally designated PMBT providers must be fully integrated into their trust's corporate and clinical governance arrangements.

There is an expectation that practitioners will participate in continuous professional development and networking.

See also NHS England service standards for the primary malignant bone tumours service.

## **4. Key Service Outcomes**

**Outcomes:**

- overall survival between 60 and 90%
- local recurrence rate of around 5-10%
- amputation rate of around 10-15%
- accuracy of diagnosis
- adherence to agreed pathways and shared protocols
- quality of life and long-term adverse effects of treatment
- patients' satisfaction with service and patient information

**5. Location of Provider Premises**

The service is delivered across England by five designated centres which provide cover across all regions in England for the national caseload. Designated services are based at:

- Oxford University Hospitals NHS Trust
- Royal National Orthopaedic Hospital NHS Trust
- The Newcastle Upon Tyne Hospitals NHS Foundation Trust
- The Robert Jones and Agnes Hunt Orthopaedic Hospital NHS Foundation Trust
- The Royal Orthopaedic Hospital NHS Foundation Trust

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