# Guide to Sarcoma for Allied Health Professionals

Sarcoma UK
The bone & soft tissue cancer charity

sarcoma.org.uk

# About this guide

This guide is for allied health professionals who treat sarcoma patients but whose majority caseload is not sarcoma. It's particularly relevant for those who work in the community with sarcoma patients who have been discharged from a specialist centre.

Already equipped with a wide range of skills and expertise in managing patients in the community, this resource simply aims to help AHPs feel confident in working with a population of patients who face many challenges throughout their pathway.

Allied health professionals are vital in the sarcoma patient pathway, particularly after surgery. It's the extent and complexity of the surgery required when treating sarcoma which often sets it apart from other cancers.

Alongside their specialist roles, they also often provide a support system for patients and carers, using a holistic approach to provide individualised care.

The guide will provide an overview of sarcoma, the clinical pathways, treatment options and post-treatment impact. It also signposts to further sources of support for allied health professionals, as well as for patients and carers.

You might want to skip between sections, use the booklet for reference or read only the most relevant sections.

**If you have any questions about anything you read in this booklet, you can contact our HCP Support Line team:**

**Phone: 0808 801 0401**
**Email: supportline@sarcoma.org.uk**
**Text: 07860 058830**

# Contents

# The basics

# Allied Health Professionals

There are a wide range of Allied Health Professionals in the UK, regulated by their respective professional bodies. Their roles are diverse and each has a role to play in the sarcoma patient pathway. They include, but are not limited to:

Arts therapists

Chiropodists and Podiatrists

Dietitians

Occupational Therapists

Orthoptists

Orthotists

Osteopaths

Paramedics

Physiotherapists

Play therapists

Practitioner psychologists

Prothetists

Radiographers

Speech and Language Therapists

Our further resources section signposts to more information about AHPs.



# What is sarcoma?

Sarcomas are uncommon cancers that can affect any part of the body.

## Soft tissue sarcoma

Soft tissue sarcomas develop in supporting or connective tissue such as muscle, nerves, tendons, blood vessels and fatty and fibrous tissues.

They commonly affect the arms, legs and trunk and also occur in:

- Stomach and intestines (GIST)
- Abdomen
- Head and neck
- Female reproductive system

## Bone sarcoma

Bone sarcoma is a cancer that starts in the bone. It can affect any bone in the body but is most commonly found in the limbs.

## GIST

Gastrointestinal stromal tumours are soft tissue sarcomas but have a very different treatment pathway to other sub-types.

They develop in the gastrointestinal tract and are most commonly found in the stomach and the small bowel. However, they can occur anywhere along the GI tract.

**3**
**main types**

**100+**
**sub-types**

**1.4%**
**of all cancer diagnoses in the UK are sarcomas**

# Most common types of soft tissue sarcoma

## Angiosarcoma

- Develops from the cells that make up the walls of blood vessels
- Most commonly found in the skin, breast, liver, spleen, and in the deep tissues
- Angiosarcoma in the skin is often found on the face and scalp. In some rare cases, it can occur in the heart
- Most common in older adults

## Fibroblastic sarcoma

- Develops in the fibrous tissues within the body
- Most commonly found in the limbs, skin and in the trunk
- Most common in those over 65 years old

## Kaposi's sarcoma

- Caused by the HHV-8 virus
- Occurs when a person has a compromised immune system which can't control the virus
- 94% of Kaposi's sarcoma cases in the UK are caused by both HHV-8 and HIV infection, the remaining 6% are caused by HHV-8 with no HIV present
- Affects the skin, mouth and occasionally the internal organs

## Leiomyosarcoma

- Develops in the smooth muscle cells
- Most commonly found in the limbs, behind the abdomen (retroperitoneum), gastrointestinal tract and the female reproductive system
- Most common in adults

## Liposarcoma

- Develops from the fat cells found all over the body
- Most commonly found in the trunk, limbs and in the retroperitoneum
- Most common in adults

## Malignant peripheral nerve sheath tumour

- Develops in the cells that cover nerves
- Can occur anywhere throughout the body
- Most common in adults

## Rhabdomyosarcoma

- Develops in the skeletal or voluntary muscles of the body
- Can occur anywhere throughout the body but most commonly found in the head and neck, as well as the abdomen
- More common in children than adults

## Synovial sarcoma

- Develops in cells around joints and tendons
- Can occur anywhere throughout the body but often near the knee
- Most common in young adults

# Most common types of bone sarcoma

## Chondrosarcoma

- Develops in the cartilage cells
- Most commonly found in the upper arm, pelvis and thigh bone
- Most common in those over 40 years old

## Chordoma

- Chordomas are most commonly found in the spine or the skull. The majority are found in the sacrum
- Most common in adults in their 40s and 50s

## Ewing's sarcoma

- Most commonly found in the pelvis, thigh bone and shin bone
- In rare cases, it can develop in the soft tissue around the bone – this is called soft tissue sarcoma. It can also start beneath the skin
- Most common in teenagers and young adults

## Osteosarcoma

- Most commonly found in the femur and tibia/fibula or humerus
- Most common in teenagers and young people but can also affect older adults

# Benign conditions managed by sarcoma MDTs

Patients may also be referred to sarcoma specialist centres with benign (non-cancerous) tumours. These can include:

## Desmoid Fibromatosis

- Most commonly found on the torso but can also appear on the arms, legs, head and neck
- Can be locally aggressive but does not metastasise
- Most common in those between 20 and 50 years old

## Giant cell tumours of the bone

- Most common in the long bones found in the arms and legs
- Can be locally aggressive and metastasis only occurs in around 2–3% of cases
- Most common in those between 20 and 30 years old

For more information on these benign tumours, you can download our Desmoid-type Fibromatosis booklet and our Giant Cell Tumours of the Bone factsheet on our website.

# Why is sarcoma different?

### Sarcoma can affect people of any age

Sarcoma can affect a person at any stage of life, from pre-natal to old age.

### Sarcoma can occur in any part of the body

Sarcoma is a cancer of the connective tissues and can appear anywhere in the body. Symptoms can vary depending on size and location.

### There is no 'one size fits all' standard treatment pathway

- Sarcomas can affect any part of the body
- Surgical resections can be very functionally limiting
- Patients may require complex reconstructions
- Patients may have multimodal treatment
- No two sarcoma patients are ever the same

It is therefore not possible for sarcoma rehabilitation to follow standard protocols. However, there are rehabilitation guidelines for some of the sarcoma surgeries.

### Treatments can be very radical

Surgery for sarcoma often requires the removal of body structures – soft tissue, muscle, nerve and bone. This can:

- Cause a huge physical change
- Cause long-term disability
- Have a significant impact on swallowing and communication in the short and long term
- Severely impact quality of life and everyday functioning, including the ability to work, socialise, drive
- Significantly impact psychological wellbeing
- Make post-surgery rehabilitation much longer

Both patients and carers will need to adjust both physically and emotionally, as well as coping with ongoing adjuvant cancer treatments.

> **"Sarcoma patients have complex surgeries involving excision of bone and soft tissues that will leave them with long term functional difficulties."**
>
> **Geraint Davies, Macmillan Specialist Practitioner**

### Chemotherapy won't be an effective treatment for every patient

For sarcomas where chemotherapy is a recommended treatment, it's important to be aware that patient responsiveness can't be predicted and chemotherapy may be stopped if the side effects are too toxic.

### Children, teenagers and young adults have a different treatment pathway

Ewing's sarcoma and osteosarcoma commonly occur in younger patients and treatment usually lasts a lot longer, around 18 months. It's important to be aware that an often overlooked side effect of treatment for this age range is the impact on their fertility.

### Sarcoma is very complex in how it acts and has a high recurrence rate

- You should be aware of and watch out for the signs and symptoms of recurrence, which can be found on p. 44
- Community therapists play a vital role in identifying late effects and signs of recurrence

### Sarcoma can metastasise in the lungs

The lungs are a common site of metastatic disease.

# “No two sarcoma patients are ever the same.”

Lynsey Green, Clinical Specialist Sarcoma Physiotherapist

## There is often a significant psychological impact to diagnosis and treatment

- Sarcoma patients often struggle with acceptance over being diagnosed with a rare cancer
- If amputation is needed, patients have to deal with both the cancer diagnosis and the loss of a limb
- Amputees and some limb salvage patients often struggle with loss of function and sensation, as well as an altered body image
- Side effects of treatment such as hair loss, physical changes after surgery, changes to facial cosmesis, and long term changes to communication and swallowing can also affect body image and have a significant psychological impact
- Sarcoma is unpredictable and has a high rate of recurrence, causing a lot of stress and uncertainty, even after successful treatment

## Patients and carers often feel very isolated

Most people have never heard of sarcoma. When they or their loved one receives a diagnosis, they have no frame of reference.

# Clinical pathways

# Clinical presentations

Sarcoma is a rare cancer and on average, a GP may only see one sarcoma in their career. Therefore, these patients often experience delays at the diagnosis stage. This can sometimes lead to a lack of trust in healthcare professionals and building rapport with this group of patients is vital.

GPs are advised to look for the following clinical presentations:

### Soft Tissue Sarcoma

- Lump more than 5cm in size
- Lump increasing in size
- Deep to the fascia

### Bone Sarcoma

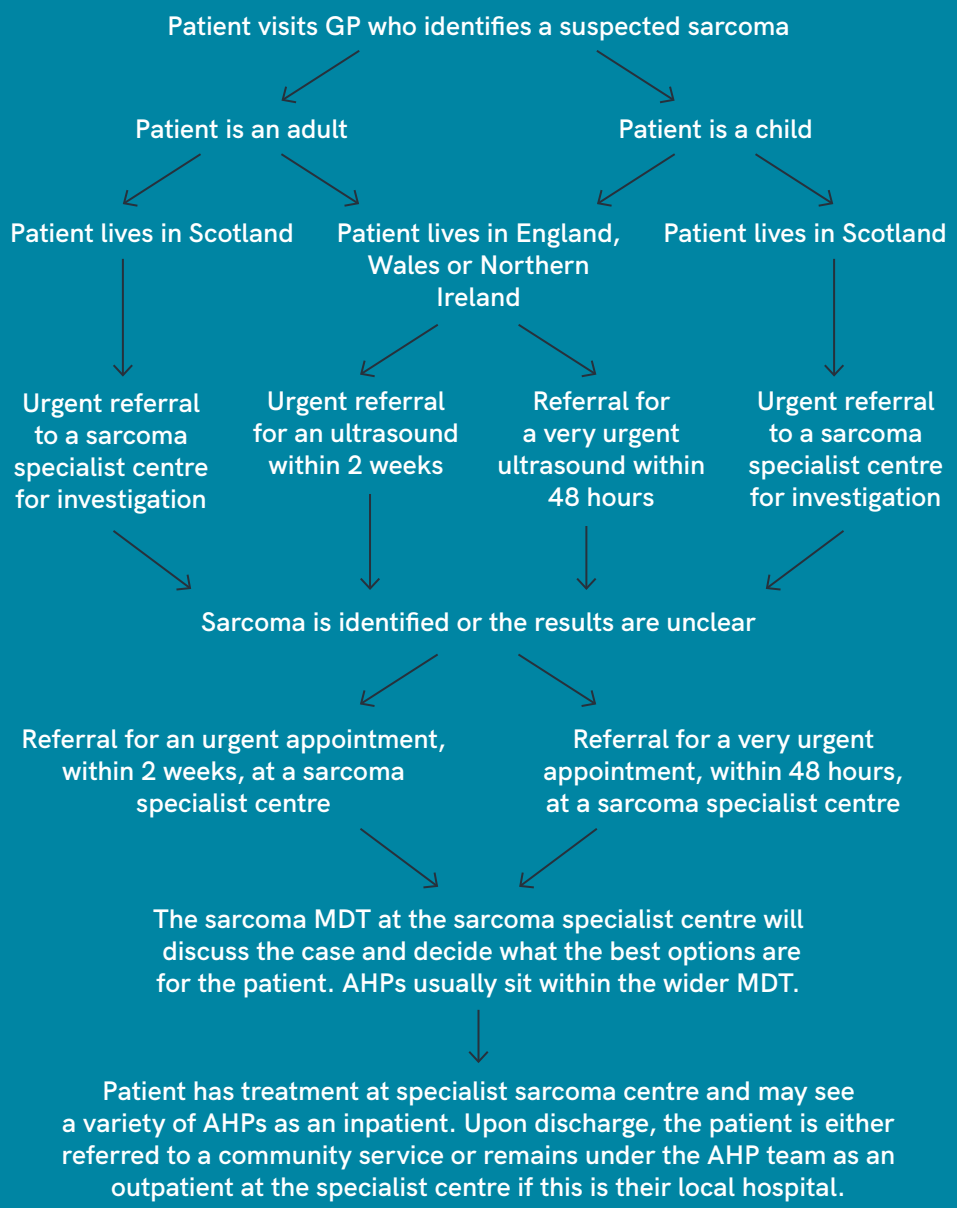
Unexplained bone tenderness which is:

- Persistent
- Increasing
- Non-mechanical
- Nocturnal or at rest

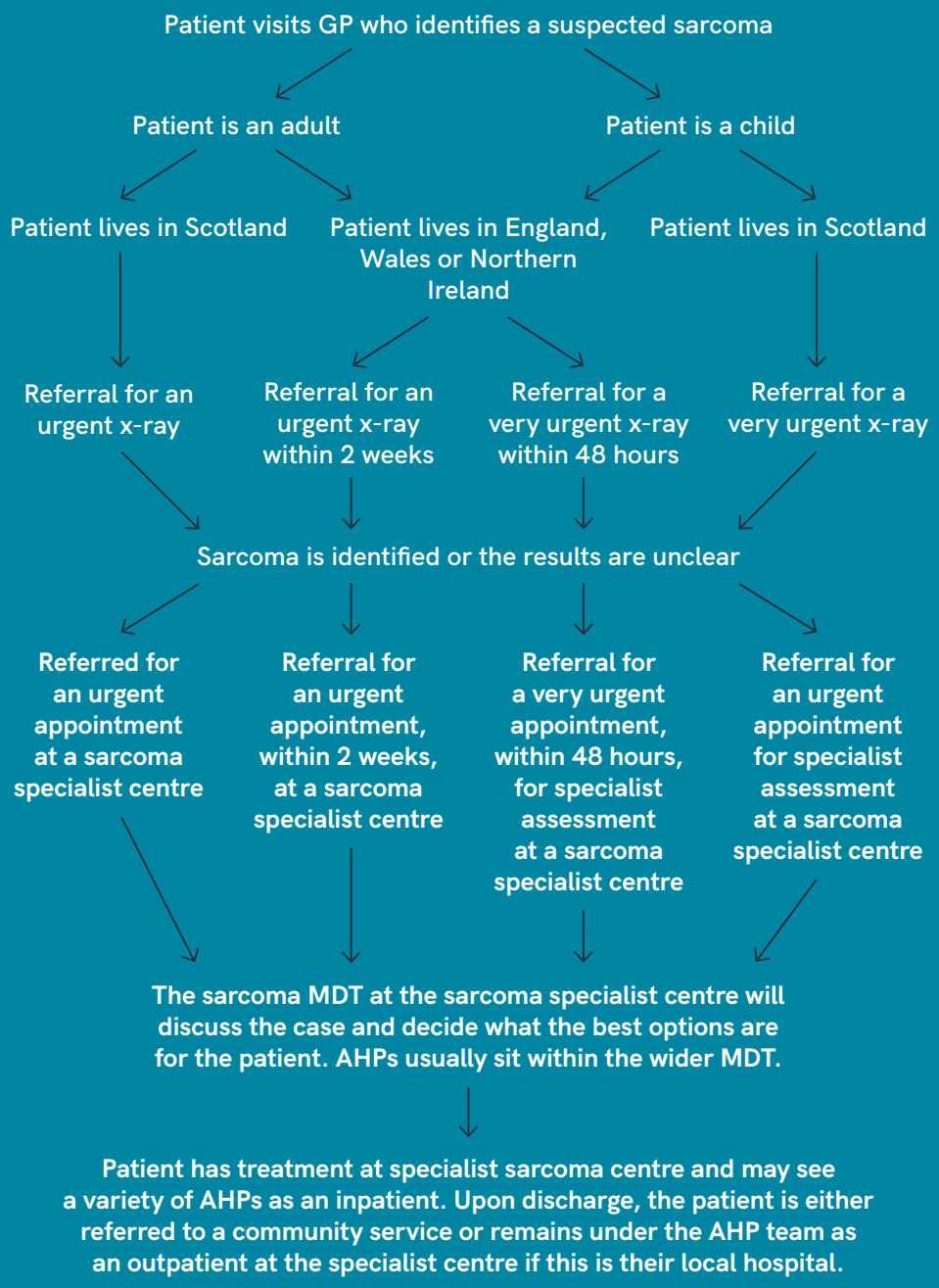
Once a sarcoma has been identified, patients should follow a standardised pathway and treatment can occur quickly.

You can download diagnostic toolkits for GPs on our website.

## Diagnostic Pathway: Soft Tissue Sarcoma



# Diagnostic Pathway: Bone Sarcoma






# Grading

It's important to know the stage and grade of your patient's cancer. These descriptions can provide a common language which is useful when discussing your patient's case with other health professionals who are overseeing your patient's care.

### Low-grade

The cancer cells are slow-growing, look quite similar to normal cells, are less aggressive, and are less likely to spread

### Intermediate-grade

The cancer cells are growing slightly faster and look more abnormal

### High-grade

The cancer cells are fast growing, look very abnormal, are more aggressive and are more likely to spread

Early AHP involvement in providing prehabilitation can make a significant difference to patient outcomes. Encouraging and supporting patients to take a proactive approach to their treatment is key, as is managing expectations. Our further resources section signposts to further information about prehabilitation.

# Staging

The stage of cancer is measured by how much it has grown or spread. This can be seen on the results of your patient's tests and scans. The results from a biopsy can tell what grade the cancer is.

### Stage 1
The cancer is low grade, small (less than 5cm) and has not spread to other parts of the body

### Stage 2
The cancer is of any grade, usually larger than stage one but has not spread to other parts of the body

### Stage 3
A high grade cancer that has not spread to other parts of the body

### Stage 4
A cancer of any grade or size that has spread to any other part of the body

# Staging for gynaecological sarcoma

Unlike other sarcomas, the staging system used to see how far a gynaecological sarcoma has spread is called the FIGO system. It is specifically used to stage cancers of the cervix, uterus, ovary, vagina, and vulva.

### Stage 1a
The cancer is small (less than 5cm) and has not spread to other parts of the body

### Stage 1b
The cancer is larger than 5cm and has not spread to other parts of the body

### Stage 2a
The cancer is of any grade, usually larger than stage one and has spread to other parts of the female reproductive system

### Stage 2b
The cancer has spread to tissues in the pelvis other than those of the female reproductive system

### Stage 3a
A high grade cancer that has spread to tissues in the abdomen in one site

### Stage 3b
A high grade cancer that had spread to tissues in the abdomen in more than one site

### Stage 3c
The cancer has spread to the lymph nodes

### Stage 4a
A cancer of any grade or size that has spread to the bladder and/or rectum (back passage)

### Stage 4b
A cancer of any grade or size that has spread to a distant part of the body from the original tumour, e.g. the lungs

# Specialist Centres

Sarcoma services are organised and delivered through designated sarcoma specialist centres, which host sarcoma multidisciplinary teams (MDTs). It is vital that patients are referred to these centres as these teams have specialist knowledge and expertise in treating sarcoma. There are 17 centres in the UK.

Patients may need to travel large distances to receive treatment at specialist centres. To try to prevent this as much as possible, patients can receive certain treatments, such as chemotherapy and radiotherapy, at a closer hospital. There are satellite hub centres linked with the specialist centres.

Any treatments at these centres will be in line with the recommendations of the MDT at the specialist centre. However, many patients place a lot of trust in specialist services and can be reluctant to be referred back to any service in the local area.

Also, patients may still have to travel significant distances to receive treatment and it's vital that transition of care across treatment locations is coherent. Collaboration with the wider community is also crucial, for example, with a patient's local social services MDT.

**England**

**Bone and soft tissue specialist centres:**

1. North of England Bone and Soft Tissue Tumour Service/Newcastle Teaching Hospitals NHS Foundation Trust

2. Greater Manchester and Oswestry Sarcoma Service (GMOSS)/Robert Jones and Agnes Hunt Orthopaedic Hospital Oswestry/Manchester Royal Infirmary/The Christie NHS Foundation Trust Manchester

3. Royal Orthopaedic Hospital NHS Foundation Trust Birmingham

4. OXSARC – The Oxford Sarcoma Service/Nuffield Orthopaedic Centre

5. The London Sarcoma Service/University College London Hospitals NHS Foundation Trust/Royal National Orthopaedic Hospital NHS Trust

**Soft tissue specialist centres:**

6. Liverpool Sarcoma Service/Royal Liverpool & Broadgreen University Hospitals Trust

7. Leeds Regional Sarcoma Service/Leeds Teaching Hospitals NHS Foundation Trust. Bone sarcomas referred to Royal Orthopaedic Hospital Birmingham.

8. Sheffield Sarcoma Unit/Sheffield Teaching Hospitals NHS Foundation Trust. Bone sarcomas referred to Royal Orthopaedic Hospital, Birmingham & Royal National Orthopaedic Hospital NHS Trust, Stanmore.

9. The Midland Abdominal & Retroperitoneal Sarcoma Unit/University Hospitals Birmingham NHS Foundation Trust.

10. East Midlands Sarcoma Service/Nottingham University Hospital NHS Trust/University Hospitals of Leicester NHS Trust. Bone sarcomas referred to Royal Orthopaedic Hospital Birmingham.

11. Bristol Sarcoma Service/North Bristol NHS Trust. Bone sarcomas are referred to the Oxford Sarcoma Service.

12. Plymouth Hospitals NHS Trust. Exeter Sarcoma Service

13. Exeter Sarcoma Service/Royal Devon & Exeter NHS Foundation Trust.

14. The Royal Marsden Hospital. The London Sarcoma Service. Bone sarcomas referred to Royal National Orthopaedic Hospital NHS Trust.

**Wales**

(2) North Wales. All soft tissue and bone sarcomas are referred to Greater Manchester and Oswestry Sarcoma Service (GMOSS).

15. South Wales Sarcoma Multi-Disciplinary Team. Bone sarcomas referred to Royal Orthopaedic Hospital Birmingham.

**Northern Ireland**

16. Sarcoma Multi-Disciplinary Team. Bone sarcomas and soft tissue sarcomas are diagnosed and treated across a number of hospitals including Musgrave Park Hospital Belfast, Ulster Hospital Dundonald, and Belfast City Hospital.

**Scotland**

17. Scottish Sarcoma Network Soft tissue and bone sarcomas are treated at the following regional centres:

Aberdeen
Dundee
Edinburgh
Glasgow
Inverness



**"It's vital that transition of care across treatment locations is coherent."**

**Charlotte Harvey-Wright, Advanced Practice Radiographer in Sarcoma**


**You can find out more information about the specialist centres and sarcoma MDTs in the NICE guidelines. Please see the references section on p.53.**

# Sarcoma MDTs

Sarcoma multidisciplinary teams (MDTs) are responsible for the diagnostic pathway and treatment of sarcoma patients. For site specific sarcomas, for example, head and neck or gynaecological, there should be a formal relationship between the sarcoma MDT and the site specific MDT.

### The Core Team

- Surgeon
- Radiologist
- Pathologist
- Medical and/or clinical oncologist
- Clinical Nurse Specialist/ key worker

### The Extended MDT

- Specialist allied health professionals
- Specialist nurse (including palliative care nurses and appropriately trained ward staff)
- Affiliated medical or clinical oncologist from a linked cancer centre
- Affiliated diagnostic service clinicians
- Other professionals including: orthopaedic, thoracic, plastic, head and neck, gynaecological, GI and vascular surgeons
- Paediatric oncologist

Also undertaking a vital role are palliative care specialists and support staff.

The Clinical Nurse Specialist is usually the keyworker and should be your first port of call for any questions you have about your patient.



# Treatment

# Aims of treatment

Understanding the intent of treatment will enable you to support patients and carers in the best possible way. Also, sometimes a general prognosis is good to know for your own emotional wellbeing.

## Curative

Treatment to destroy the cancer.

## Palliative

Also known as supportive care.

Treatment, care and support designed to help control symptoms and improve quality of life. It can include:

- Pain management
- Emotional, spiritual and psychological support
- Social care
- Support for family and friends

It's important to consider whether rehabilitation would be beneficial, with adaptive rehabilitation often being very important to palliative patients.

Palliative is often used to mean treatment for people who can't be cured. However, a patient that is palliative may live for many years and function to a high level.

## End of Life

Treatment, care and support for those nearing the end of their life. Timeframes can be difficult to predict and some patients will receive this care for longer than others. It aims to help patients live as comfortably as possible in the time they have left. It can include:

- Symptom management
- Emotional support for them and loved ones
- Expressing needs and wishes
- Practical matters such as making a will

Hospices can facilitate rehabilitation for palliative patients, as well as end of life care. Hospice UK have produced downloadable resources on this topic, available on their website. Our further resources section signposts to these.



# Soft Tissue Sarcoma

## Surgery

Surgery is the standard treatment for most patients. The surgeon will remove the tumour and will aim to take out an area of normal tissue around it.

Limb salvage will be attempted with amputation occurring if it is the best or only means to achieve adequate margins.

## Radiotherapy

**Low Grade**
Most patients won't need radiotherapy.

**Intermediate or High Grade**
For most patients, radiotherapy either before or after surgery is considered to be the standard treatment.

**Before surgery**
This can help to reduce the size of the sarcoma to aid surgical excision and may also reduce the risk of recurrence.

**After surgery**
The aim is to kill off any local cancer cells which remain in the area of the tumour.

**Radiotherapy as the only treatment method**
This is unusual but in a small number of cases where surgery isn't possible, it can lead to a full or part reduction in symptoms. However, the likelihood of recurrence is high.

## Isolated limb perfusion

This is a treatment in which the blood circulation of the extremity is isolated and local treatment with chemotherapy is applied in the isolated area.

Where available, this may be used before surgery to reduce the size of a tumour and may allow for limb preservation.

## Proton Beam Therapy

Proton beam therapy is a form of radiotherapy which uses high energy proton beams instead of x-ray radiation beams. This means that it can deliver a more targeted use of radiotherapy than x-ray radiotherapy.

There are two main groups who would benefit from PBT rather than x-ray radiotherapy:

- Children and young people as radiotherapy can sometimes damage their developing organs.
- Adults with sarcoma in parts of the body which are hard to treat, as there is a smaller radiation dose to surrounding tissues. This would include areas such as the spine, the base of the skull, the head and neck region and the pelvis.

## Clinical trials

If the treatments above have not been successful, patients may be considered for clinical trials. A clinical trial can sometimes be used as part of primary treatment if a suitable trial is open which the patient is eligible to join.

## Chemotherapy

The benefits of chemotherapy to treat soft tissue sarcomas hasn't been proven. It may be considered:

- On higher risk tumours or on sarcoma subtypes which are more responsive to this treatment
- Before surgery where surgery wouldn't be straightforward

**Metastatic disease**

In most cases of metastatic soft tissue sarcoma.

- First-line chemotherapy: either doxorubicin or combined doxorubicin and ifosfamide.
- Second-line chemotherapy: Trabectedin or combined gemcitabine and docetaxel.

For more information on Proton Beam therapy, you can download our factsheet on our website.



**"Specific to sarcoma patients is the need for a high level of individualised care."**

**Charlotte Harvey-Wright, Advanced Practice Radiographer in Sarcoma**

## Management of local recurrence

In most cases, where possible, surgery will be undertaken to regain local control. Radiotherapy can also be used if not used previously. Amputation may be needed in selected cases.

## Management of lung metastases

Potential treatments include surgery, radiofrequency or microwave ablation, or stereotactic ablative radiotherapy.

## Management of metastases elsewhere in the body

In most cases, metastases outside the lungs will be treated with systemic treatment. In selected cases surgery, radiofrequency ablation, cryotherapy or radiotherapy may be considered for limited metastatic disease to prolong remission or reduce symptoms.

Our further resources section signposts to the Cancer Research UK website, where you can learn more about individual chemotherapy drugs.

## Metastatic disease

**Treatment**

In almost all cases, the treatment intention is palliative. Treatment is complex and individualised, dependent on symptoms and the potential side effects of treatment. For patients without significant symptoms, disease stabilisation is the aim, to prolong good quality of life. For many patients, standard supportive care with symptom control alone is often the most appropriate option.

**Treatment options**

A combination of treatment strategies may be used, especially for patients with a prolonged disease course. These may be used to control symptoms, reduce pain or try to prolong the remission period. These may include:

- Systematic treatment
- Surgery
- Radiotherapy
- Microwave or radiofrequency ablation
- Medication related to pain management and other complications such as bone metastases

## Clinical trials

If other treatment options have not been successful, patients may be considered for clinical trials.

**"The MDT all work closely together to manage these patients as their needs are highly complex."**

**Florence Cook, Head and Neck Dietitian**



# Gastrointestinal Stromal Tumours

## Surgery

Surgery is usually the standard treatment, with complete removal of the tumour.

## Drug interventions

If surgery would have a major functional impact, the drug imatinib is standard therapy prior to surgery to reduce the tumour size.

This is standard therapy for 3 years for patients with a significant risk of relapse, provided their tumour is not likely to be resistant to therapy.

## Metastatic disease

**Surgery**

For residual metastatic disease, surgery may be an option. Surgery of limited progression along with continual imatinib may be a palliative option.

Selected patients with limited liver metastases may have surgery or radiofrequency ablation.

**Where surgery isn't an option**

- Imatinib is the standard first-line treatment
- Sunitinib is the second-line treatment
- Regorafenib is the third-line treatment

The patient will progress to the next line of treatment if there is confirmed progression or intolerance to the drug.

Patients with metastatic GIST failing all three standard agents should be considered for participation in clinical trials.

Radiotherapy can sometimes be used in lower doses to relieve symptoms.

# Desmoid Fibromatosis

## Watchful waiting

This is the first line approach to treatment and can also be known as ‘watch and wait’, ‘active monitoring’ or ‘active observation’.

**Duration**

Patients are usually on a watch and wait programme for 1–2 years. This can vary depending on individuals. It ensures effective treatment can be held in reserve for when patients most need it. If the patient’s condition stabilises or regresses this will continue.

## Surgery

Surgery is not used routinely due to potential high recurrence except in very certain circumstances, for example, for abdominal wall tumours, or if there is a risk to surrounding structures such as arteries/veins.

## Radiotherapy

When surgery is not an option and active management is required, moderate dose definitive radiotherapy has also been shown to provide adequate local control in a majority of progressive patients.

## Drug interventions

Systemic treatment options comprise anti-hormonal therapies such as tamoxifen or toremifene, non-steroidal anti-inflammatory drugs (NSAIDs), tyrosine kinase inhibitors (TKIs), and “low-dose” or conventional chemotherapeutic regimens including liposomal doxorubicin.

## Exceptions

When the tumour is located near to a critical structure that may pose significant problems to the patient’s life, an earlier decision towards an active therapy may be taken due to the potentially higher risk of morbidity prior to disease stabilisation. The type of further treatment is generally guided according to the anatomical site and the decision should be made with the patients in a stepwise approach.



# Bone Sarcoma

## Osteosarcoma

**Surgery and Chemotherapy**

Curative treatment for high-grade osteosarcoma consists of surgery and chemotherapy. For low grade osteosarcoma, only surgical intervention is usually required.

The aim of surgery is to safely remove the whole tumour whilst preserving as much function as possible. Most patients with extremity tumours are candidates for limb salvage.

Chemotherapy treatment commonly takes 6–9 months, comprising 10 weeks of neo-adjuvant therapy, surgery and adjuvant chemotherapy. The most accepted regimen is:

- Induction therapy with MAP (high-dose methotrexate, doxorubicin and cisplatin)
- If not tolerated, patients under 40 years old may be given doxorubicin and cisplatin alone
- Mifamirtide may be offered to patients without metastases after surgery

**Recurrent and metastatic disease**

Treatment for locally recurrent or metastatic osteosarcoma is surgery, if possible. Pulmonary metastatectomy can lead to long term survival if all metastases can be completely removed.

There is however a role for chemotherapy and radiotherapy within a palliative care context for symptom management.

## Chondrosarcoma

The standard treatment is surgery to remove the tumour. Chemotherapy may have a role in some subtypes. Radiotherapy is only used as definitive treatment if surgery isn’t possible.

## Ewing’s sarcoma

Systematic treatment with chemotherapy is standard. When treatment aims to be curative, all of the pre-chemotherapy volume should be treated with surgery, radiotherapy or both.

Pre-operative radiotherapy may be advantageous.

**Recurrent and metastatic disease**
Patients should be considered for clinical trials.

## Giant Cell Tumour of the Bone

Surgery is usually the first treatment option. It's sometimes treated with the biological therapy denosumab.

**Recurrent and metastatic disease**
This could involve further surgery or denosumab treatment.

## Chordoma

Where possible, the standard treatment is surgery to remove the tumour. Proton beam therapy may be used after surgery for some patients, particularly those with high sacral tumours.

Surgical excision of tumours of the skull base or cervical spine aims to remove as much tumour as possible, whilst preserving neurological function and therefore quality of life.

**Recurrent and metastatic disease**
Metastases are rare but local recurrence is common and difficult to cure. Treatment for local recurrence may include surgery and/or radiation therapy and/or systematic treatment. Molecular targeted agents may be effective.

**"As sarcoma is rare, other health professionals as well as the patient might not have clear ideas of the treatment pathway."**

**Edwina Hudspeth Stevenson, Occupational Therapist**

# Post treatment

# Risks and side effects

## Surgery

**Changes to limb function**
This is most likely where muscle, bloods vessels or parts of bone are removed along with the tumour, there is damage to nerves, or where patients have had spinal surgery.

Surgery can be very functionally limiting, resulting in restricted or altered range of movement post surgery. Gait aids or adaptive footwear may need to be considered. Patients may also require complex reconstructions or amputation surgery.

**Amputation of limb**
This is most likely with bone sarcomas or where it's the only surgical option to achieve adequate margins.

If amputation of a limb occurs, patients may be fitted with a prosthetic limb. Patients will be required to undergo a lengthy fitting and rehabilitation processes after surgery to learn how to use a prosthetic.

Patients may require the use of aids and equipment to manage everyday activities and complex wound management and healing after surgery may be needed.

**Changes to bladder, bowel and sexual function**
This is most likely where GISTs are removed in the sacrum or bowel or where patients have had spinal surgery. Also, retroperitoneal patients, depending on the size and location of their tumour, may have part or all of their organs removed, such as the kidney, colon (bowel), pancreas, spleen or bladder

Patients may need a colostomy or urostomy following surgery.

The Royal National Orthopaedic Hospital has produced Rehabilitation Guidelines for sarcoma patients. Our further resources section signposts to these.

**"Patients requiring amputation can feel anger, depression, and/or anxiety and can go through the stages of grief around loss of limb/function. Other effects can include phantom limb pain and sensation, amongst others."**

**Val Jacques-Robinson, Clinical Prosthetist**

**Changes to speech and swallowing**
This is most likely where surgery is extensive or with head and neck patients.

Patients often need Speech and Language Therapy intervention to address difficulties with swallowing and communicating after treatment. This may include swallowing exercises and strategies, and dietary modification.

**Changes to diet**
Patients often need modification to dietary textures, as well as nutritional support, including management of enteral feeding when required.

For example, GIST patients who have had part or all of their stomach removed will receive specialist support from a dietitian.

As sarcoma patients often require extensive rehabilitation, you'll get to know them well. Be aware of the emotional impact on yourself if your patient is diagnosed with recurrence or metastatic disease or if they die. Make sure you discuss it with your team.

## Radiotherapy

Side effects are usually limited to the treatment site. Depending on the location of the radiotherapy field and the type and intensity, they may include:

- Tiredness
- Loss of hair in the treatment area
- A dry mouth
- Breathing problems
- Infertility
- Low sex drive
- Erectile dysfunction
- Soreness and pain
- Bowel changes
- Bladder inflammation
- Radiation fibrosis
- Changes to the skin, including dryness, irritation, redness, swelling and/or blistering and peeling. Skin side effects are often worse where two skin surfaces meet so may need longer to heal
- Difficulty swallowing, including aspiration risk, voice changes and speech changes
- Secondary cancer, including radiation induced sarcoma many years after initial treatment
- The need for vaginal dilators, which may be recommended for use to minimise the risk of vaginal stenosis. Their infrequent use may be recommended indefinitely

## Proton Beam Therapy

The side effects will depend on the body part being treated and the tumour size. Changes to the skin may include:

- Dryness
- Irritation
- Redness
- Swelling
- Blistering or peeling

## Oral anticancer targeted drugs

Common side effects include:

### Imatinib

- Fatigue
- Swollen ankles
- Nausea
- Diarrhoea
- Itchy rash
- Puffy eyes

### Sunitinib

- Fatigue
- Mouth ulcers
- High blood pressure
- Skin rash and soreness

### Regorafenib

- Fatigue
- Loss of appetite
- Pain
- Diarrhoea
- Weight loss

## Chemotherapy

Common side effects include:

- Increased risk of getting an infection
- Breathlessness and looking pale
- Feeling sick or vomiting
- Bruising, bleeding gums or nosebleeds
- Tiredness and fatigue
- Loss of appetite
- Diarrhoea
- Hair loss
- Risk of cardiac toxicity with certain drugs

The effects of chemotherapy and/or radiotherapy can impact on a patient's ability to engage with rehabilitation. For amputees, rehabilitation with a prosthesis may have to be delayed until afterwards, due to the physical demands of using a prosthetic limb.

# Function

Sarcoma derives from the connective tissues of the body and therefore the mere presence of a tumour can directly impact on function, as can subsequent treatment.

This has implications not only on mobility but also on falls, ability to carry out activities of daily living, to drive, work and participate in normal social and leisure activities. The impact on psychosocial wellbeing can be significant. A holistic and MDT approach to care is therefore essential.

## Surgery

Surgery can involve resection of muscle, nerves, bones, skin and/or blood vessels. It may also include repair of some of these structures, with or without plastic reconstruction.

It is therefore important to be aware of any local protocols or rehabilitation precautions. The treatment a patient receives will determine expectations for recovery and prescribed AHP intervention.

## Radiotherapy

Treatments such as radiotherapy can have an impact on function in the short and longer term, due to the risk of lymphoedema and radiation induced fibrosis, for example. Patients should be prepared for this.

This is especially relevant when treatment is of a large area of muscle or crosses a joint, as this can have long term implications on mobility, activities of daily living and quality of life.

**"Treatments such as radiotherapy can have an impact on function in the short and longer term so people should be prepared for this. Managing expectations is key."**

**Lucy Dean, Specialist Sarcoma Physiotherapist**



# Lymphoedema

Lymphoedema is a swelling caused by a build-up of fluid in the tissues under the skin. Sarcoma patients could be at increased risk of developing it because some treatments can damage the lymphatic system. The causes are irreversible and there is currently no cure.

Patients may be at risk of developing lymphoedema if they've had:

- Lymph nodes removed as part of surgery
- Radiotherapy to an area of the body where lymph vessels and lymph nodes are located

## Signs and symptoms

The most common symptom of lymphoedema is a swelling in the arm or leg caused by a build-up of fluid under the skin. Other symptoms that can occur in the part of the body affected include:

- A feeling of heaviness or tightness
- Restricted range of movement
- Aching or discomfort
- Recurring infections
- Hardening and thickening of the skin called fibrosis

Lymphoedema may develop several weeks, months or even years after patients have had treatment. Swelling is normal following surgery or radiotherapy but if it hasn't gone down within 6–8 weeks, you should contact the patient's Clinical Nurse Specialist, who can advise you on next steps.

You can download our factsheet and z-card on lymphoedema on our website.

# Late effects

Sarcoma patients can develop late effects of treatment, those which begin during or shortly after treatment and continue for six months or more. Alternatively, they can become permanent and some can begin months or even years after treatment. Interventions for late effects can be preventative, remedial, or both. Late effects can include:

## Cancer-related cognitive impairment

This side effect is often missed and misdiagnosed in patients. It is mostly associated with chemotherapy and radiotherapy treatments.

## Communication impairments and difficulty swallowing (dysphagia)

Communication impairments and difficulty swallowing (dysphagia).

Where required, patients are provided with prophylactic swallowing exercises prior to radiotherapy and surgery, as well as exercises for communication and swallowing following these treatments.

## Fatigue, insomnia and trouble sleeping

The biggest side effect of cancer and its treatment is fatigue. There is evidence that being physically active can help manage fatigue. Fatigue management strategies are also important.

Graded activity and exercise, energy conservation, structured psychological support, relaxation and structured sleep as well as cognitive rehabilitation can all be helpful. This will need to be individualised to the person that you are treating.

## Heart and kidney problems

Some chemotherapy patients experience long term heart and kidney problems which may need to be dealt with years after the treatment has ended.

## Infertility

Patients receiving chemotherapy can have infertility issues and where appropriate, patients will be referred to a fertility specialist prior to commencing treatment.

## Lymphoedema

Surgical patients are at increased risk of developing lymphoedema. Early education about the risks can empower patients to take action to reduce the risk of lymphoedema developing.

## Peripheral neuropathy

Chemotherapy may cause peripheral neuropathy. Some people will experience numbness, tingling and/or pain in the hands and/or feet and this can affect mobility and balance. It may affect some people long term and in these cases, an MDT approach may be required to help manage it.

## Radiation fibrosis

This is scar tissue that forms as a result of damage from radiation therapy and can affect mobility.

## Scar management

Some patients will have scars which might not heal properly and that have adhered to underlying tissue. Depending on the location, this can cause issues with mobility, gait and posture. Advice on scar management is important.

# Signs of recurrence

A recurrence is a tumour that comes back in the same place. The signs to look out for are:

### Soft Tissue Sarcoma

- New lumps/swellings in/around the site of the previous tumour removal, including on/around a scar
- New pain that cannot be attributed to injury

### GIST

- Changes to bowel habits
- Bloating
- Pain
- Obvious mass

### Bone

- A decrease in movement of joints and/or reduced mobility
- New bone pain that is more prominent at night

### Gynaecological

- New bleeding
- Bloating
- Pain

**If you have any concerns, you should discuss these urgently with the team at the specialist centre.**

"Community therapists and local hospitals play a pivotal role in providing an early warning safety net for patients and identifying signs of recurrence."

Charlotte Betteridge, Children's Occupational Therapy Team Lead

# Questions to consider

## Treatment: Do you know?

- The name of the bone/soft tissue that has been removed
- The technicalities of surgery performed and post-operative instructions and restrictions
- Details of any adjuvant treatments
- Future management plans
- Where the patient is in their treatment cycle
- What side effects the patient is experiencing
- What matters to the patient
- If the patient has accessed all available forms of support

This information should be available in the handover notes provided by the therapy team at the specialist centre and/or by asking your patient. Further sources of support for patients can be found on p. 51.

### Have you?

- Undertaken your own assessment
- Adopted a teamwork approach with the patient
- Managed expectations
- Set realistic goals

### Are your goals for the patient?

- Holistic
- Flexible
- Progressive
- Individualised

### What is your role in the patient's pathway?

- "It's important to understand that they have often spent a lot of time in hospital and we are only one very small part of their journey."

  Donna McGuigan, Clinical Lead Orthotist



# Checklist

### Prior to seeing a post-op surgical patient, you should find out:

- [ ] What structures have been resected and what functional impact is this likely to have?
- [ ] What reconstruction have they had to close the wound? What limitations might this mean? What interventions do you need to promote or avoid?
- [ ] How much are the consultants happy for the patient to do?
- [ ] What other treatment has the patient had? Neo-adjuvant Radiotherapy? Chemotherapy? How might this effect their function, healing, exercise tolerance etc?

### During follow up:

- [ ] Where was the patient's primary tumour and any metastatic disease? Knowing this will help you to identify signs of recurrence
- [ ] What treatment have they had? What are the likely long term implications? Possible late effects?
- [ ] How is the patient managing psychologically?

### Patients having chemotherapy:

- [ ] What chemotherapy regime are they having and what are the common side effects? How might this impact their rehabilitation?
- [ ] Are they having any other treatments e.g. surgery?
- [ ] What prehabilitation would be beneficial?
- [ ] Do they have restrictions?

### Patients having radiotherapy:

- [ ] Where is the radiotherapy field?
- [ ] What structures is this likely to effect?
- [ ] What interventions can you suggest to reduce these effects?
- [ ] What position does the patient need to be in for their radiotherapy?
- [ ] Can the patient achieve this and maintain it for several minutes?

# Further information

# Important contacts for you

## Clinical Nurse Specialist

Every patient is allocated a key worker and this is usually the sarcoma specialist CNS, who is an invaluable source of information for AHPs about their patient. The CNS should be your first port of call for any questions you have about your patient. The CNS:

- Provides advice, support and continuity
- Is the point of contact for patients and carers
- Has specialist sarcoma knowledge
- Acts as a patient advocate
- Facilitates the patient pathway

An AHP may be the key worker in long-term follow-up.

## Therapy team at the specialist centre

The therapy team will be familiar with the patient's case and have specialist expertise about sarcoma.

## The multidisciplinary team

Other key contacts are the members of the patient's MDT, including their surgical consultant and oncologist.

## Sarcoma UK Support Line

The Support Line is staffed by HCPs who can answer any questions you may have:

**Phone:** 0808 801 0401
**Email:** supportline@sarcoma.org.uk
**Text:** 07860 058830



# Signposting patients and carers

If you feel that your patient needs extra support, you can signpost them to:

Their clinical nurse specialist or doctor, who may also be able to refer them for additional support.

The Sarcoma UK Support Line is here for everyone who is affected by sarcoma. The Support Line is staffed by HCPs:

**Phone:** 0808 801 0401
**Email:** supportline@sarcoma.org.uk
**Text:** 07860 058830

There are sarcoma support groups around the UK, as well as online support groups. Details can be found on our website or by contacting our Support Line.

Sarcoma UK has information resources for patients which can be downloaded from our website. Hard copies are also available.

Other charities which support sarcoma patients include the Bone Cancer Research Trust and GIST Cancer UK.

Other charities which aren't sarcoma specific but provide valuable support and services include Maggie's, Macmillan, Citizens Advice, Cancer Haircare Charity, MOVE and Shine Cancer Support.
The Teenage Cancer Trust provides support for young people.
CLIC Sargent help children, young people and their families.

Patients can download our 'Understanding Rehabilitation and Life after treatment' booklet.

# Further resources

You may find the following online resources helpful:

**For more information about sarcoma:**

**British Sarcoma Group**
Clinical guidelines and recommendations for sarcoma can be found on the British Sarcoma Group website:
britishsarcomagroup.org.uk/treating-sarcoma/guidelines/

**Sarcoma UK**
Other Sarcoma UK information resources are available in print and/or are downloadable from our website:
sarcoma.org.uk/support-information/patient-guides

**For more information about the sarcoma specialist centres:**

**Sarcoma UK**
sarcoma.org.uk/health-professionals/sarcoma-specialist-centres

**For more information about treatment, prehabilitation and rehabilitation:**

**Cancer Research UK**
cancerresearchuk.org/

**Hospice UK**
Rehabilitative Palliative Care online resources:
hospiceuk.org/what-we-offer/clinical-and-care-support/rehabilitative-palliative-care/resources-for-rehabilitative-palliative-care

**Royal National Orthopaedic Hospital**
Rehabilitation Guidelines (Sarcoma):
rnoh.nhs.uk/services/rehabilitation-guidelines

**Macmillan Cancer Support**
macmillan.org.uk/cancer-information-and-support/impacts-of-cancer

**Macmillan, Prehabilitation for people with cancer (2019)**
macmillan.org.uk/assets/prehabilitation-guidance-for-people-with-cancer.pdf

**For more information about lymphoedema:**

**British Lymphology Society**
thebls.com/documents-library

**For more information about the roles of individual Allied Health Professionals:**

**Health and Care Professions Council**
hcpc-uk.org

**NHS**
england.nhs.uk/ahp/role

**The British Association of Play Therapists**
bapt.info

**The General Osteopathic Council**
osteopathy.org.uk/home



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## Websites

**Cancer Research UK:**
cancerresearchuk.org/about-cancer/cancer-in-general/treatment

**This guide has been written in collaboration with allied health professionals who currently work with sarcoma patients. They have generously contributed their collective clinical knowledge and experience.**

**This booklet has been produced by the Information and Support Team at Sarcoma UK.**
**It has been reviewed by allied healthcare professionals.**



If you would like to provide us with feedback on this resource, please email **feedback@sarcoma.org.uk**

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# Notes

Sarcoma UK
The bone & soft tissue cancer charity

# Sarcoma UK is a national charity that funds vital research, offers support for anyone affected by sarcoma cancer and campaigns for better treatments

@Sarcoma_UK
@Sarcoma_UK
uk.sarcoma

sarcoma.org.uk