Welcome to this NDRS training module on Sarcoma, which has been designed to help Cancer Administration staff gain a better understanding of these tumours and the terminology used by the clinical teams.

Agenda

- Sarcoma
- Summary
- Acknowledgements

This module may be paused at any time

We'll look at the some facts about bone and soft tissue Sarcomas, their diagnosis & treatment and the coding to use. Remember, this module can be paused at any time.

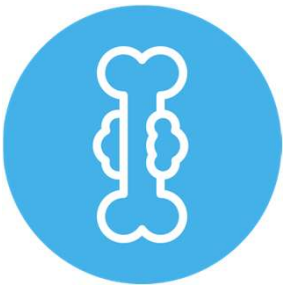
Sarcoma

In this section we will cover:

- Introduction
- Causes and Risk Factors
- Signs and Symptoms
- Anatomy & Physiology
- Regional Lymph Nodes
- Diagnosis
- Morphology
- ICD10 coding
- Grade
- Stage
- Treatment

Sarcoma - Introduction

- Rare cancer ~ 1% of all malignancies
- Arise in bone and soft tissue
  - More than 100 subtypes
- Can arise anywhere in the body

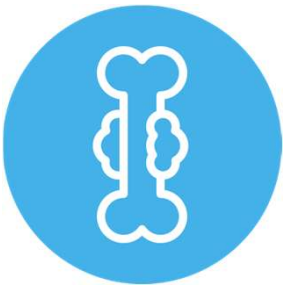


Sarcomas are rare - only around 1% of malignancies are determined to be a sarcoma, either of the bone or soft tissue. A sarcoma may arise anywhere in the body

Sarcoma – Causes & Risk Factors

The cause of connective tissue tumours is mostly unknown but there are some factors which are known to increase the risk

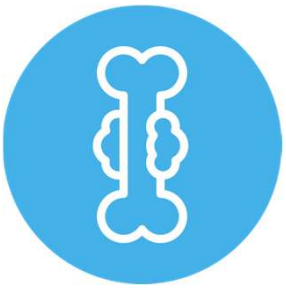
- Age
- Family history/ genetics including: Neurofibromatosis, Li Fraumeni disease, Retinoblastoma Syndrome
- Exposure to radiation
- Compromised immune system (relevant to Kaposi sarcoma)
- Other bone disease



Depending on the subtype, sarcomas can develop in people of all ages but the overall risk does increase with age – around 10% of soft tissue sarcomas are in people under the age of 30 whereas roughly 40% occur in people over the age of 65. Family history, radiation exposure and prior bone disease are also known to increase the risk of developing a sarcoma

Sarcoma – Signs & Symptoms

- Pain
- Swelling
- Tiredness
- Fever or sweats
- Weight loss
- Difficulty with movement (bone)
- Pathological fracture (bone)

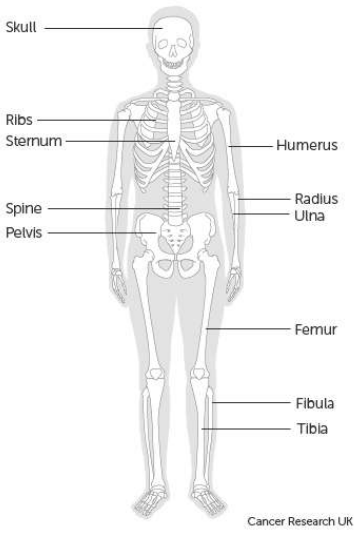


Some symptoms are specific to sarcomas of the bone, such as a pathological fracture or difficulty moving. Other signs of a possible sarcoma include pain, swelling or weight loss.

Sarcoma – Anatomy & Physiology - Bone

The adult skeleton contains 206 bones. The function of the skeletal structure is to:

- Support other tissues
- Protect vital organs
- Act as levers for the muscles to enable us to move

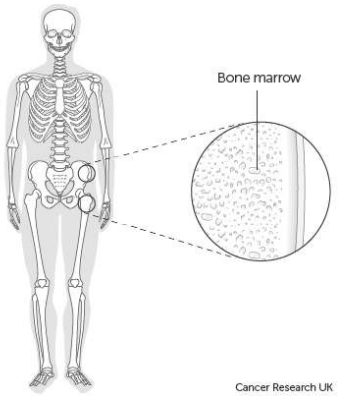


The skeleton has multiple functions. It supports other tissues such as muscle, fat and skin – it protects vital organs within the ribcage – and it provides leverage to allow the muscles to move us around.

Sarcoma – Anatomy & Physiology - Bone

Bones generally consist of 3 distinct layers:

- Periosteum: a tough membrane that covers the outside of the bone
- Cortical bone: dense, solid bone that surrounds the marrow space
- Trabecular bone: a honeycomb-like structure of bone within the marrow space. Bone marrow sits in the gaps within this structure.

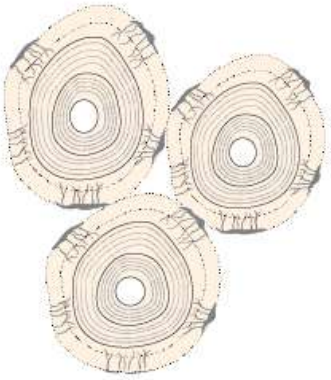


Bones are usually comprised of three layers: a tough membrane on the outside, underneath that - a layer of solid bone … and in the middle: a honeycomb structure of bone in the marrow space.

Sarcoma – Anatomy & Physiology - Bone

Bone Cells:

- Osteoblasts: these form new bone
- Osteoclasts: these break down unnecessary bone
- Osteocytes (pictured): fully formed bone cells



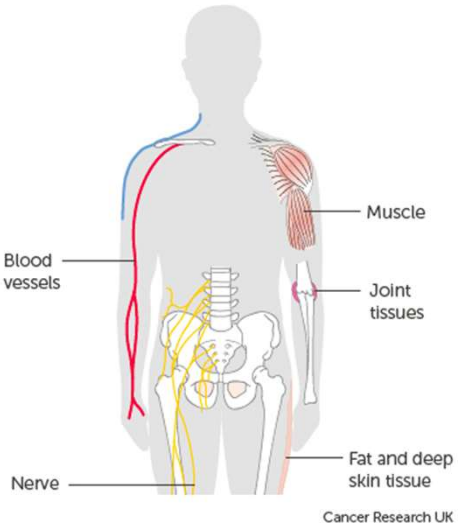
Cancer Research UK

Bone itself is formed on a framework of cartilage. This framework also incorporates minerals including calcium - which gives the bones their hardness – as well as specific types of cells. The 3 main bone cell types work together to form and maintain the skeletal structure:
If a bone injury occurs, the osteoblasts make new bone framework to repair the damage, occasionally making too much.
The osteoclasts remove any extra framework, reshaping the bone in the process. As bone forms, osteoblasts are trapped within it and become part of the hardened structure. These fully formed bone cells are known as osteocytes.

Sarcoma – Anatomy & Physiology – Soft Tissue

There are different types of soft tissue

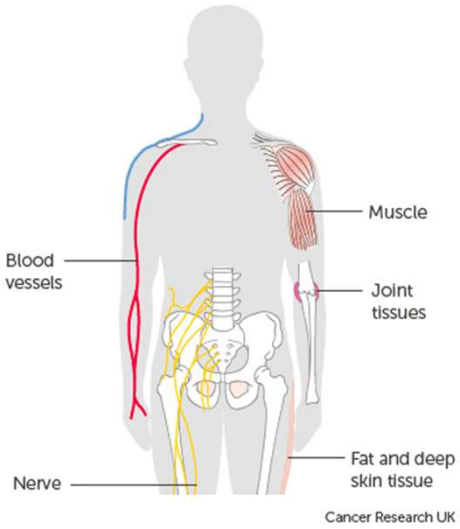
- Muscle
  - Skeletal muscle – subject to voluntary control
  - Smooth muscle – not subject to voluntary control, found in the uterus, digestive system and blood vessels
  - Cardiac muscle – the muscle that forms the heart



Soft tissue sarcomas can occur in a number of different types of body tissue, one of which is muscle. Muscle may be of the type over which we have control, such as that found in the limbs… it may be of the smooth type which functions without voluntary control and can be found in the digestive system and blood vessels … or it may be the muscle that forms the heart.

Sarcoma – Anatomy & Physiology – Soft Tissue

- Fat & deep skin tissues
- Blood vessels
- Nerves
- Tendons and ligaments
- Tissues around the joints

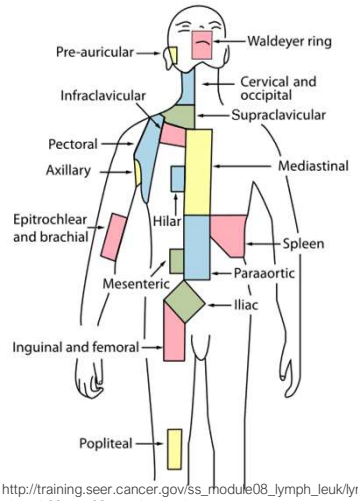


Other tissues in which sarcomas may form include the deep layers of the skin, fatty tissue, nerve tissue and the tissues around the joints.

Sarcoma – Regional Lymph Nodes

The regional lymph nodes for sarcomas are those appropriate to the site of the primary tumour

Regional lymph node involvement is rare for sarcoma



http://training.seer.cancer.gov/ss_module08_lymph_leuk/lymph_unit02_sec02_reg_lns.html

It's rare for sarcomas to include regional lymph node involvement … but when it does occur the regional lymph nodes will be appropriate to the site of the primary tumour.

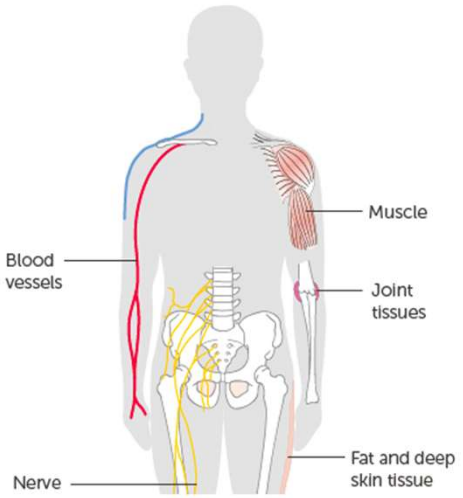
Sarcoma - Diagnosis

- Physical Examination
- Imaging:
  - X – Ray
  - Ultrasound Scan
  - CT Scan
  - MRI Scan (pictured)
  - PET Scan
- Blood tests
- Biopsy

Diagnosis of a sarcoma usually involves physical examination, one or more types of imaging and potentially blood tests and/or a biopsy.

Sarcoma – Morphology – Soft Tissue

Tissue type the Sarcoma most resembles	Tumour type
Fat	Liposarcoma
Smooth muscle	Leiomyosarcoma
Skeletal muscle	Rhabdomyosarcoma
Peripheral nerves	Neurofibrosarcoma Malignant peripheral nerve sheath tumour
Fibrous tissue	Fibrosarcoma
Blood vessels	Angiosarcoma
Lymphatic vessels	Lymphangioma
Synovial tissue	Synovial Sarcoma



The morphology of a sarcoma will depend on the type of cell they most resemble which is not necessarily the cell type in which the tumour has arisen. There are more than a hundred different subtypes. Clues to the type of sarcoma cell are often in the first part of the name. For instance: a Lipo- prefix means the sarcoma most closely resembles fat cells, Fibro- fibrous tissue and Angio- blood vessels. Specific morphology would normally be detailed on a path report where a biopsy or surgical removal has taken place.

Sarcoma – Morphology – Bone & Soft Tissue

Ewing Sarcoma

- Can develop in soft tissue as well as bone
- Most common in teenagers
- Morphology coded to M9260/3

Ewing sarcomas can arise in either bone or soft tissue and occur mainly in teenagers

Sarcoma – Morphology – Bone

Osteosarcoma

The most common type of bone tumour with several sub-types. These are characterised by the production of bony substance (Osteoid). They may occur in patients of all ages but are the most common primary bone tumour in teenagers and young adults. Refer to the pathology report for the exact type. Morphologies include:

- Osteosarcoma, NOS – M9180/3
- Osteosarcoma, conventional - M9186/3
- Osteosarcoma, low grade – M9187/3
- Osteosarcoma, parosteal – M9192/3
- Osteosarcoma, periosteal – M9193/3
- Osteosarcoma, secondary – M9184/3
- Osteosarcoma, small cell – M9185/3
- Osteosarcoma, surface, high grade – M9194/3
- Osteosarcoma, telangiectatic – M9183/3

The most common type of bone sarcoma is an osteosarcoma, of which there are a number of sub-types including those shown here. Osteosarcomas are a common primary sarcoma in teenagers and young adults.

Sarcoma – Morphology – Bone

Chondrosarcoma

These are characterised by the formation of tumour cartilage. They are most common in the 30 – 60 age group. Refer to the pathology report for exact type. Morphologies include:

- Chondrosarcoma, NOS – M9220/3
- Clear cell chondrosarcoma – M9242/3
- Dedifferentiated chondrosarcoma – M9243/3
- Mesenchymal chondrosarcoma - M9240/3
- Periosteal chondrosarcoma – M9221/3

Chondrosarcomas occur in cartilage and are more common in the over 30s.

Sarcoma – ICD10 coding – Bone (includes articular cartilage)

Malignant neoplasm of bone and articular cartilage of limbs – C40.*

- Scapula and long bones of upper limb – C40.0
- Short bones of upper limb (includes bones in wrist and hand) – C40.1
- Long bones of lower limb – C40.2
- Short bones of lower limb (includes bones in foot) – C40.3
- Overlapping lesion of bone and articular cartilage of limbs – C40.8
- Bone and articular cartilage of limb, unspecified – C40.9

ICD10 coding of bone sarcomas is largely topographical – the code tells you where the tumour is. In this instance, the C40 codes relate to specific bones and joints in the arms and legs…

Sarcoma – ICD10 coding – Bone (includes articular cartilage)

Malignant neoplasm of bone and articular cartilage of other and unspecified sites – C41.*

- Bones of skull and face – C41.0
- Mandible – C41.1
- Vertebral column – C41.2
- Ribs, sternum and clavicle – C41.2
- Pelvic bones, sacrum and coccyx – C41.4
- Overlapping lesion of bone and articular cartilage – C41.8
- Bone and articular cartilage, unspecified – C41.9

… while other bones and joints are covered in C41. It should be noted that both C40 and C41 include sarcomas of the cartilage in the skeletal joints

Sarcoma – ICD10 coding – Soft Tissue

Kaposi Sarcoma – C46.*

- Kaposi Sarcoma of skin – C46.0
- Kaposi Sarcoma of soft tissue – C46.1
- Kaposi Sarcoma of palate – C46.2
- Kaposi Sarcoma of lymph nodes – C46.3
- Kaposi Sarcoma of other sites – C46.7
- Kaposi Sarcoma of multiple organs – C46.8
- Kaposi Sarcoma, unspecified – C46.9

The ICD10 codes for soft tissue sarcomas may also indicate the type of sarcoma. C46 is specific to Kaposi sarcoma…

Sarcoma – ICD10 coding – Soft Tissue

Malignant neoplasm of other connective & soft tissue (includes: blood vessel, bursa, cartilage, fascia, fat, ligament (except uterine), lymphatic vessel, muscle, synovia, tendon (sheath):

- Connective and soft tissue of head, face and neck (excludes orbit) – C49.0
- Connective and soft tissue of upper limb including shoulder – C49.1
- Connective and soft tissue of lower limb including hip – C49.2
- Connective and soft tissue of thorax (excludes breast, heart, mediastinum & thymus) – C49.3
- Connective and soft tissue of abdomen – C49.4
- Connective and soft tissue of pelvis – C49.5
- Connective and soft tissue of trunk, unspecified – C49.6
- Overlapping lesion of connective and soft tissue – C49.8
- Connective and soft tissue, unspecified – C49.9

… while other sarcomas are often coded to C49, depending on the location of the primary tumour…

Sarcoma – ICD10 coding – Soft Tissue

Exclusions to C49 include:

- Articular cartilage (ie; joints) – C40.* & C41.* (where * denotes the specific location)
- Connective tissue of breast – C50.*
- Cartilage of larynx – C32.3
- Cartilage of nose – C30.0
- Peripheral nerves & autonomic nervous systems – C47.*
- Peritoneum – C48.*
- Retroperitoneum – C48.0

… It should be noted that there are exceptions to this. As already mentioned, articular cartilage is coded to C40 or C41, while others are coded as shown here…

Sarcoma – ICD10 coding – Soft Tissue

- Malignant neoplasm of orbit – C69.6
- Malignant neoplasm of heart & mediastinum – C38.* (where * denotes the specific location)
- Malignant neoplasm of thymus – C37X
- Malignant neoplasm of other and unspecified female genital organs, broad ligament – C57.1
- Malignant neoplasm of other and unspecified female genital organs, round ligament – C57.2
- Malignant neoplasm of other and unspecified female genital organs, parametrium, includes uterine ligament NOS – C57.3

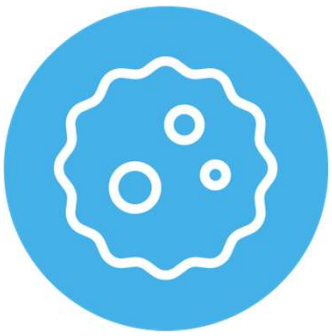
… and here. As always, an asterix indicates more specific coding with the final digit … while a final X indicates a top level only code.

Sarcoma – Grade - Bone

Grade for bone sarcomas indicates the biological behaviour of a tumour and contributes to the calculation of stage. The grading system is:

- Low grade (includes grade 1)
- High grade (includes grade 2 and 3)

- Some morphological types are associated with a specific grade - Ewing sarcoma is always grade 3



As with many grading systems, the closer the resemblance of tumour cells to normal, healthy cells, the lower the number. For bone sarcomas, the numeric grade is translated into Low and High grade tumours.

Sarcoma – Grade – Soft tissue Sarcoma

The grading system used for soft tissue sarcomas in the UK is the French Federation of Cancer Centres Sarcoma Group (Fédération Nationale des Centres de Lutte Contre Le Cance - FNCLCC) which is derived following assessment of:

- Mitotic rate – the rate at which cancer cells reproduce, scored from 1 (low reproduction rate) to 3 (high reproduction rate)
- Presence of necrosis – the presence and proportion of dying tissue in the sarcoma, scored from 0 (no necrotic tissue) to 2 (50% or more necrotic tissue)
- Differentiation – how closely the sarcoma cells resemble normal tissue in appearance and functionality, scored from 1 (similar to normal cells) to 3 (very abnormal cells)

- Scores are added together to give the grade.
  - Grade 1 means a total score of 2 or 3 (sometimes called LOW grade)
  - Grade 2 means a total score of 4 or 5 (grade 2 or 3 are sometimes called HIGH grade)
  - Grade 3 means a total score of 6 or more

Grading of Soft Tissue Sarcomas relies not only on the resemblance to normal tissue but also on the rate of reproduction and the presence of any dead tissue within the tumour. Each of these factors is given a score which is then added to the other factors to assign the Grade.

Sarcoma – Stage

- Unless another staging system is specified it's important that we receive the Final Pre-Treatment and/or Final Integrated Stage as follows:
  - For diagnosis dates up to 31st December 2025 use UICC TNM v8
  - For diagnosis dates from 1st January 2026 use UICC TNM v9
- Please note that the TNM version must be accurately recorded – if you are unable to amend the version on your cancer data management system, please refer to your line manager
- If, after 1st January 2026, your cancer data management system has not been amended to include TNM v9 please record the TNM v9 stage and add the following statement to the Primary Diagnosis Subsidiary Comment field:
  - Patient staged using TNM9 not TNM8 as per CR2070

Unless otherwise specified, stageable sarcomas must be staged using the appropriate UICC TNM version for the purposes of COSD.

Sarcoma – Stage

- For details on recording stage, please see the NDRS training module KPI-TNM Staging 101, available on this link: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Stage should be recorded for all stageable sarcomas. Staging data sheets for sarcomas may be downloaded from: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets

For details on recording Stage please refer to the NDRS training module KPI-TNM Staging 101, which explains the staging process…

Sarcoma – Stage - Bone

- Grade of the tumour is important to assign group stage
- T stage is based on size & location of the primary tumour and presence of discontinuous tumours in the primary site: Appendicular Skeleton, Trunk, Skull & Facial Bones; Spine; Pelvis
- Sarcomas of the spine or pelvis must be staged using the T, N & M components although no group stage system has been determined for them
- The relevant UICC TNM version must be used as follows:
  - For diagnosis dates up to 31st December 2025 use UICC TNM v8
  - For diagnosis dates from 1st January 2026 use UICC TNM v9

… however, with sarcomas of the bone, it should be noted that pelvic and spinal sarcomas do not have a group stage allocated to them. T, N & M elements of the stage should still be recorded.

Sarcoma – Stage – Soft Tissue

Soft Tissue Sarcoma is staged as follows:

- For diagnosis dates up to 31st December 2025 use UICC TNM v8
- For diagnosis dates from 1st January 2026 use UICC TNM v9

- Please note that the TNM version must be accurately recorded – if you are unable to amend the version on your cancer data management system, please refer to your line manager
- If, after 1st January 2026, your cancer data management system has not been amended to include TNM v9 please record the TNM v9 stage and add the following statement to the Primary Diagnosis Subsidiary Comment field:
  - Patient staged using TNM9 not TNM8 as per CR2070

Similarly, most soft tissue Sarcomas are stageable at the level of T, N & M…

Sarcoma – Stage – Soft Tissue

- T stage is assigned based in size and location, with different classifications depending on location: Extremity and Superficial Trunk; Retroperitoneum; Head & Neck; Thoracic and Abdominal Viscera
- Sarcomas of the Head and Neck or Thoracic and Abdominal Viscera must be staged using the T, N & M components although no group stage system has been determined for them
- GIST and uterine Sarcomas are staged with their respective site-specific systems
- Stage must be recorded for all stageable cases

… although, as with bone, some don't have stage group classifications. T, N & M stage would still need to be recorded for all stageable sarcomas using the appropriate UICC TNM version. There **are** some exceptions: GISTs would be staged using the appropriate TNM staging system for the GIST location and uterine sarcomas would be staged using the relevant FIGO staging system

Sarcoma – Treatment – Surgery

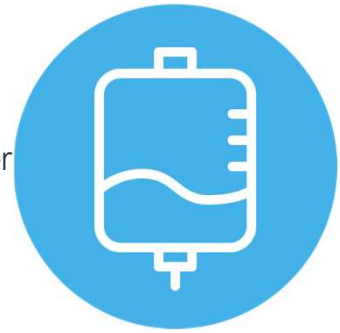
The most common treatment for sarcoma is surgery, aiming to remove all the tumour while leaving as much of the functional tissue behind as possible

- The extent of surgery necessary depends on the size and location of the tumour
- Amputation may be the most appropriate treatment in order to offer the best chance of cure, however, preservation of limbs will be achieved wherever possible

Surgery is a common treatment for sarcomas, with the extent of the surgery being dependent on the size and location of the tumour.

Sarcoma – Treatment – Chemotherapy

- Neo-adjuvant chemotherapy is commonly used for sarcomas in young people such as osteosarcoma, Ewing sarcoma and rhabdomyosarcoma. It can also be used for larger and high grade soft tissue sarcomas to shrink them prior to surgical resection
- Isolated limb perfusion (ILP) is a method of localised delivery of chemotherapy to a limb and is performed under general anaesthesia
- Palliative chemotherapy is recommended for recurrent tumours and metastatic sarcoma



Chemotherapy is sometimes used to shrink a tumour prior to surgery. It may also be offered as a palliative treatment.

Sarcoma – Treatment – Radiotherapy

Radiotherapy is not usually the first treatment given to patients with sarcoma

- Neo-adjuvant radiotherapy is sometimes considered for patients with tumours that are borderline operable
- Adjuvant radiotherapy may be given to patients with intermediate or high grade tumours as it is proven to reduce the risk of local recurrence
- Radiotherapy can be offered as a palliative treatment to treat symptoms and alleviate pain

Radiotherapy may be offered neo-adjuvantly if the tumour is borderline operable. It may also be offered either as an adjuvant treatment to reduce the risk of recurrence or as a palliative treatment

Summary

In summary…

Summary

- Risk factors for sarcomas include age, genetic factors, prior radiation exposure or prior bone disease

Sarcomas are more likely in specific age groups (depending on the type)… where particular genetic mutations have occurred … or where there exists a history of bone disease

Summary

- Risk factors for sarcomas include age, genetic factors, prior radiation exposure or prior bone disease
- Signs and symptoms may include pain, swelling, weight loss or pathological fracture

Bone sarcomas may present with difficulty moving or fracture caused by a tumour in the bone. Sarcomas of either type may present with pain or weight loss

Summary

- Risk factors for sarcomas include age, genetic factors, prior radiation exposure or prior bone disease
- Signs and symptoms may include pain, swelling, weight loss or pathological fracture
- Investigations usually include physical examination and imaging and may also include blood tests or a biopsy

Diagnostic investigations normally include a physical examination and some form of imaging. Blood tests or a biopsy may also be needed

Summary

- Risk factors for sarcomas include age, genetic factors, prior radiation exposure or prior bone disease
- Signs and symptoms may include pain, swelling, weight loss or pathological fracture
- Investigations usually include physical examination and imaging and may also include blood tests or a biopsy
- The majority of sarcomas are considered stageable. Record the appropriate UICC TNM version for sarcoma or, for specified locations, the location- or site-specific stage must be used

Most sarcomas are stageable. Where a sarcoma is considered stageable, the relevant sarcoma- or location-specific staging must be recorded

Summary

- Risk factors for sarcomas include age, genetic factors, prior radiation exposure or prior bone disease
- Signs and symptoms may include pain, swelling, weight loss or pathological fracture
- Investigations usually include physical examination and imaging and may also include blood tests or a biopsy
- The majority of sarcomas are considered stageable. Record the appropriate UICC TNM version for sarcoma or, for specified locations, the location- or site-specific stage must be used
- Treatments may include surgery, chemotherapy, radiotherapy depending on the particular features and location of the tumour as well as the fitness of the patient

While surgery is the most common treatment for sarcoma, chemotherapy or radiotherapy may also be offered.

Summary

- Additional guidance on recording COSD data including morphology, topography, staging and recording a diagnosis can be found at: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Staging data sheets can also be downloaded from the NDRS website for clinical use: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets

- Staging data sheets are listed by ICD 10 code / description as applicable.
- For sarcoma these include:
  - C38.1-2, C47-C49 – Soft Tissue
  - C40-C41 – Bone
  - C69.6 – Sarcoma of Orbit
  - Rhabdomyosarcoma and ST Sarcomas

Additional training modules as well as Staging sheets for clinical use may be downloaded from the NDRS website

Summary

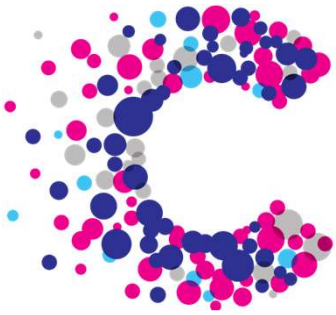
- If in any doubt as to whether you should be recording a diagnosis, please refer to the latest COSD User Guide, Appendices A & B
- For guidance on the required staging system, please refer to the latest COSD User Guide, Appendix E

- https://digital.nhs.uk/ndrs/data/data-sets/cosd#downloads

Do please remember, guidance **is** available on our website. You can download the COSD User Guide by clicking on this link and selecting the COSD version appropriate to your trust.

Acknowledgements

Many thanks to Cancer Research UK for the use of their images within this training module.



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We'd like to thank Cancer Research UK for the use of their images within this training module.

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