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# PRIMARY BONE CANCER

ANGIOSARCOMA OF THE BONE




**BONE  
CANCER**  
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UNTIL THERE'S A CURE

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# CONTENTS



- What is it?
  - Who does it affect?
  - Symptoms
  - Types of Angiosarcoma of the Bone
  - Cause and Risk Factors
  - Diagnosis
  - Treatment
  - Follow-up Care
  - Rehabilitation and Support
- 

An angiosarcoma is a soft-tissue tumour starting in cells which line the blood vessels. As it develops in the blood vessel lining, it can arise anywhere in the body – including the bone. This factsheet will focus on angiosarcoma of the bone – which accounts for less than 1% of all primary bone cancers.

## WHAT IS IT?

Angiosarcomas are very rare tumours that are able to present in any area of the body – usually in the soft-tissues. Angiosarcomas are most commonly reported in the breasts, heart, skin, liver and bones. Angiosarcomas are able to arise in various locations in the body because they develop in cells known as ‘**endothelial cells**’, which form the lining of the **blood vessels**. When an endothelial cell of the blood vessel becomes abnormal and grows uncontrollably, an angiosarcoma can develop.

Angiosarcomas of the bone make up just **6%** of all reported cases of angiosarcoma. The bones with the most reported cases of angiosarcoma are the **thigh bone**, the **shin bone**, the **pelvis** and the **spine**.

## WHO DOES IT AFFECT?

Angiosarcomas of the bone can affect anyone, at any age. Due to this cancer being rare, and there being a low number of reported cases to review, it is difficult to confirm any trend in its occurrence.

Despite this low incidence, it is thought that this cancer presents slightly more often in males than females and most frequently occurs between **20 to 50 years of age**.

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# WHAT ARE THE SYMPTOMS?

Angiosarcomas of the bone can sometimes present with no symptoms at all in the early stages – doctors describe this as ‘**asymptomatic**’.

When symptoms do present, they depend on the size and location of the tumour. The most commonly reported symptoms of an angiosarcoma of the bone are:

## BONE PAIN

this pain may be a dull ache or a stabbing sensation

## SWELLING OR A LUMP

## FATIGUE

If the tumour is located near the spine, as well as causing back pain the tumour can put pressure on the spinal cord and nerves - leading to **neurological symptoms**. These symptoms may include numbness or weakness when using the arms or legs and a sensation of pins and needles.

Patients may not experience any of these symptoms, or may only experience a few of the ones listed.

# CAUSES AND RISK FACTORS

The cause of an angiosarcoma of the bone is unknown, and studies into the cause of this cancer are difficult as it is so rare. Although there is currently no identifiable cause, there are some conditions which may increase the likelihood of an individual developing an angiosarcoma of the bone.

These include:

- **BONE INFARCTION -**

which is the death of the bone in certain areas due to a lack of blood supply and therefore oxygen to the area.

- **PAGET'S DISEASE OF THE BONE -** this is one of the most common bone disorders and causes weakening and pain in areas of the bone

- **CHRONIC OSTEOMYELITIS -** which is an infection of the bone, usually caused by bacteria.



# DIAGNOSING ANGIOSARCOMA OF THE BONE

Further tests to confirm an angiosarcoma of the bone diagnosis include:

- A CT SCAN
- AN MRI SCAN
- A BIOPSY OF THE BONE
- BLOOD TESTS

The first step in diagnosing any primary bone cancer is a trip to the GP. Diagnosis of a suspected bone tumour usually follows a **clinical examination** and an **X-ray**. It is very common to be referred to a bone cancer specialist for a second opinion and confirmation of the diagnosis.

If an angiosarcoma of the bone is presenting, the X-ray will show an aggressive appearance indicating the requirements for further tests. A CT scan and MRI scan cannot definitively diagnose an angiosarcoma of the bone. However, they provide important information on the exact location of the tumour, the stage of the tumour and evidence of any spread of angiosarcoma to other areas of the body.

Taking a biopsy is needed to confirm the diagnosis of an angiosarcoma of the bone. This specialist procedure takes a small sample of the tumour so it can be examined under a microscope.

**Results from a biopsy can take up to two weeks to analyse but they enable doctors to confirm the presence of an angiosarcoma of the bone.**



# AN ALTERNATIVE DIAGNOSIS?

There are various health conditions that may present in a similar manner to an angiosarcoma of the bone - in terms of symptoms, signs and diagnostic appearance - but it is important that the correct diagnosis is made to ensure the treatment provided is suitable. Conditions other than angiosarcoma of the bone which may be the cause of particular symptoms are known as **'differential diagnoses'**.

Other conditions which can present in the same way as adamantinoma include:

- **METASTATIC CARCINOMA -**  
when a cancer spreads from one area of the body to form a tumour in another area of the body, the secondary tumour is known as a metastatic carcinoma. Numerous cancers develop secondary tumours in the bone
- **EPITHELIOD HEMANGIOENDOTHELIOMA -**  
a rare tumour that arises in the bone, liver and the lungs.



# TREATING ANGIOSARCOMA OF THE BONE

If the presence of an angiosarcoma of the bone is confirmed the patient will be referred to the nearest Bone Cancer Centre where the specialist medical team will design the best possible treatment plan for the individual patient.

Due to the rarity of angiosarcoma of the bone, it is difficult to develop new treatments. Currently, the most effective treatment is thought to be surgery alongside chemotherapy and radiotherapy.

## SURGERY

The most effective treatment method for angiosarcoma of the bone is the surgical removal of the tumour with '**wide surgical margins**'. This means some healthy tissue is removed alongside the tumour to ensure all the tumour cells are removed and to lower the risk of the tumour returning at a later date.

Usually an angiosarcoma of the bone can be treated with '**limb-sparing surgery**', which means the tumour is completely removed whilst preserving as much of the normal function and cosmetic appearance of the limb as possible.

Common types of limb-sparing surgery performed to treat an angiosarcoma of the bone are:

- **AN AUTOGRAFT:** the affected area of bone is removed and reconstructed using the patients' own tissue from another area of their body.
- **AN ALLOGRAFT:** donated tissue is used to reconstruct the affected area of the bone once the tumour has been removed
- **A METALLIC REPLACEMENT:** once the tumour is removed the area of damaged bone is replaced with a metal implant known as a **prosthesis**. This procedure usually requires rehabilitation therapy after surgery

On rare occasion, the angiosarcoma of the bone may have spread to other areas of the body before a treatment plan is created. In these incidences, limb-sparing surgery may not be beneficial and, in order to remove the whole tumour, an amputation of the affected limb may be required.

## RADIOTHERAPY AND CHEMOTHERAPY

If the angiosarcoma of the bone is fast growing and more likely to spread elsewhere in the body, the patient is likely to receive chemotherapy before surgery is performed. This aims to shrink the tumour allowing a more successful surgical procedure, and may help to treat any tumour cells that have already spread elsewhere in the body. Chemotherapy may also be used if the patient does not wish to undergo surgery or is not fit for surgery to take place.

Radiotherapy is most frequently used after surgery has taken place. However, if the tumour has already spread to other areas of the bone, adjacent bones or other areas of the body, radiotherapy is of little use to the patient. In some cases, radiotherapy may be given to relieve the pain that patients may be experiencing – in these cases the treatment is known as '**palliative radiotherapy**'.



## FOLLOW-UP CARE



After finishing treatment, many patients will require follow-up care.

Follow-up care at the hospital will allow healthcare professionals to keep an eye on a patient's general health and ensure the patient hasn't suffered any '**LATE EFFECTS**' from their treatment. Late effects of a patient's treatment include effects on the patient's kidney function, fertility or risk of developing a secondary cancer

Follow-up care can continue for months, or even years, and allows patients to discuss any concerns they may have with their doctor. Tests may be carried out during these appointments to ensure the patient is healthy and the cancer is not at risk of returning.

## REHABILITATION AND SUPPORT



Following treatment, many patients benefit from further support and rehabilitation services.

Rehabilitation is a form of therapy that enables patients to regain strength, tackle day-to-day activities and return to normal life as quickly as possible following a disease. These services are available both during and after treatment and include:

- **PHYSIOTHERAPISTS:** help patients return back to an active lifestyle as quickly as possible to restore strength, movement and function
- **OCCUPATIONAL THERAPISTS:** help patients to complete day-to-day activities in order to regain their independence
- **DIETICIAN:** offer advice on the most appropriate nutrition for patients during and after their treatment
- **PROSTHETISTS:** specialists who design and create prostheses following amputations to match as closely as possible to the individual patients removed limb
- **ORTHOTISTS:** specialists who provide aids for patients following surgery, such as splints or special footwear

Patients, or their family and friends, may benefit from discussing any feelings of anxiety or concerns they may have following a cancer diagnosis or treatment. Many services are available for this form of support, such as:

- **PSYCHOLOGICAL SUPPORT AND SERVICES:** psychologists will support patients through any feelings of anxiety or depression to overcome the concerns that often come with a cancer diagnosis
- **LOCAL SUPPORT GROUPS:** many support groups are organised and ran locally. It is best to ask your clinical nurse specialist for information on these local services

THE BONE CANCER RESEARCH TRUST  
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TO FIGHTING PRIMARY BONE CANCER.

OUR MISSION IS TO SAVE LIVES AND IMPROVE  
OUTCOMES FOR PEOPLE AFFECTED BY  
PRIMARY BONE CANCER THROUGH RESEARCH,  
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**WE RECEIVE NO GOVERNMENTAL FUNDING,  
SO RELY ENTIRELY ON THE SUPPORT OF THE  
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