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# PRIMARY BONE CANCER

## ADAMANTINOMA

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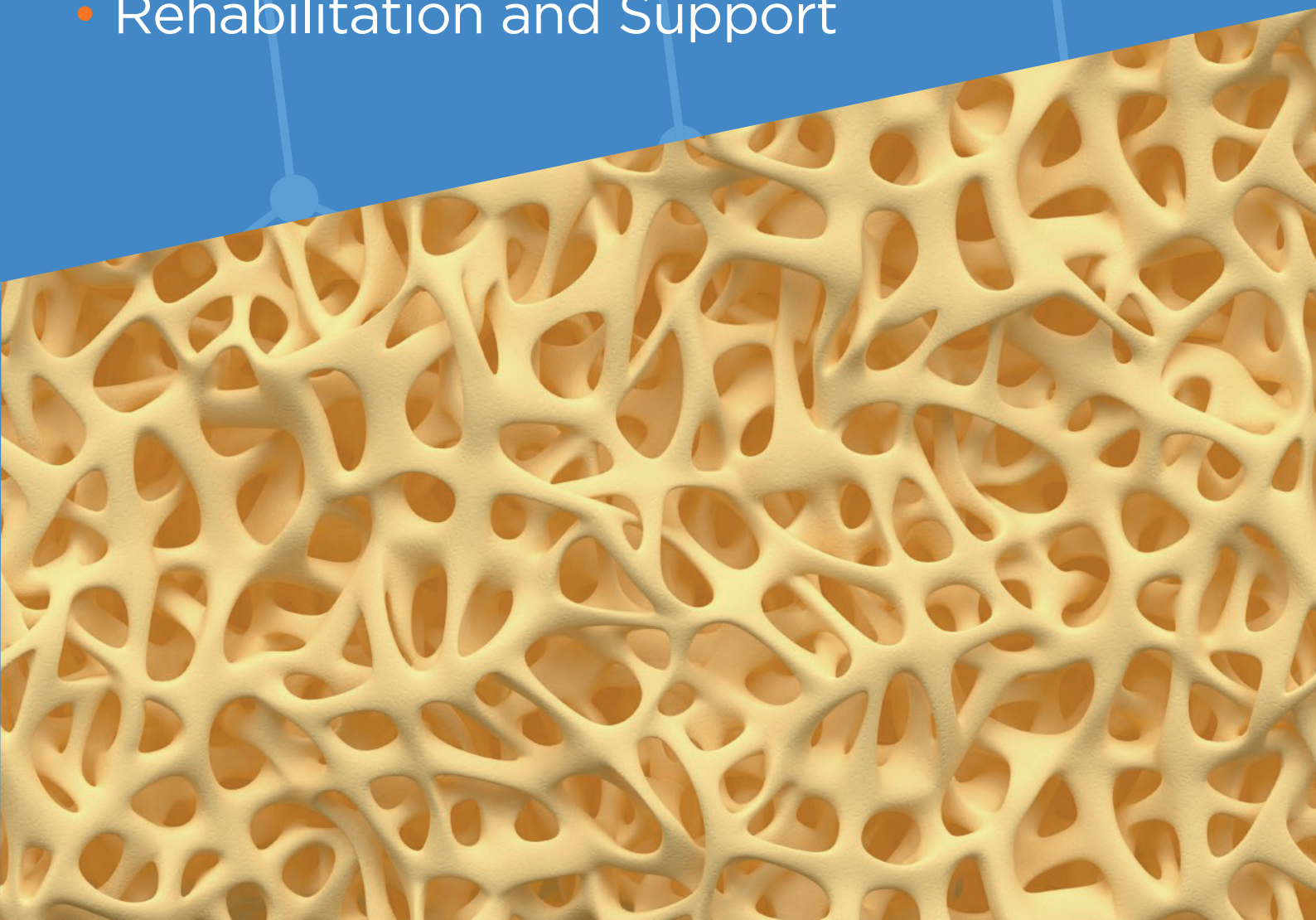
# CONTENTS





Adamantinoma accounts for less than 1% of all primary bone cancers. The most effective way to treat this rare cancer is by surgically removing the tumour.

# WHAT IS IT?

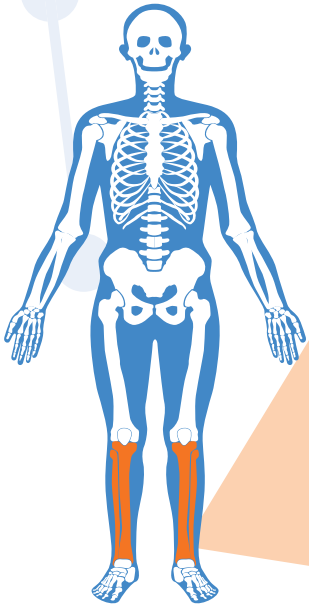
Adamantinoma is a rare form of primary bone cancer that is most commonly found in the **shin bone** and the **calf bone**, but can also be found in other bones of the body.

These tumours grow slowly and are therefore referred to as 'low-grade tumours'. Due to this slow-growth, patients often suffer from symptoms for a long period of time before going to see a doctor.

# WHO DOES IT AFFECT?

Adamantinomas can develop in anyone, at any age, but most commonly occur in the age range of **20-35 years old.**

This cancer is slightly more common in males than in females and tends to affect younger men once their bones have stopped growing - which is known as reaching **skeletal maturity.**



**MOST COMMONLY OCCURS IN THE CENTRE OF THE SHINBONE, WHICH IS KNOWN AS THE TIBIA BONE.**

# WHAT ARE THE SYMPTOMS?

Symptoms can often be **non-specific** to primary bone cancer, and some patients may not have any symptoms at all for some time. Furthermore, symptoms depend on the location of the tumour. The most commonly reported symptoms of adamantinoma are:

**INTERMITTENT BONE PAIN**

which is pain that may come and go

**AN UNEXPLAINED LIMP**

**A LUMP OR SWELLING**

**REDUCED MOVEMENT OF THE AFFECTED AREA**

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

# TYPES OF ADAMANTINOMA

By looking closely at what kind of cells are making up the tumour, doctors can class each patient's adamantinoma as one of **two** types.

## CLASSIC ADAMANTINOMA

Classic adamantinoma is the most common type of adamantinoma and is usually seen in adults over the age of around 20 years old.

## DEDIFFERENTIATED ADAMANTINOMA

which is also referred to as 'osteofibrous dysplasia-like adamantinoma'

Dedifferentiated adamantinoma is a less aggressive form of this tumour and tends to occurs in children and young people under the age of around 20 years old.

These two types can be distinguished by radiologists (who assess images of the tumour following diagnostic scans) or by pathologists (who assess the types of cells in the tumour under a microscope).

# CAUSES AND RISK FACTORS

The cause of adamantinoma is unknown. Due to the rarity of this cancer it is hard to find a possible cause from the small number of cases that have been reported.

Although there is currently no identifiable cause, approximately **60%** of adamantinoma patients have a history of a significant trauma (previous injury) to the affected bone. This may be a risk factor for developing an adamantinoma, or it may be that the tumour itself weakens the area of bone and increases the likelihood of an injury to this area. Whether a significant trauma to the bone is a risk factor for adamantinoma is still under investigation.

**60% OF ADAMANTINOMA PATIENTS HAVE A HISTORY OF A SIGNIFICANT TRAUMA**

# DIAGNOSING ADAMANTINOMA

**Further tests to confirm an adamantinoma 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.

A CT scan and MRI scan cannot definitively diagnose an adamantinoma. However, these scans do provide important information on the exact location of the tumour, the stage of the tumour and the presence of an adamantinoma having spread elsewhere in the body.

Taking a biopsy of the bone is needed to confirm the diagnosis of adamantinoma. 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, and specific type, of adamantinoma.**

# AN ALTERNATIVE DIAGNOSIS?

When diagnosing an adamantinoma, the patients' **age** and **clinical history** play a large role in being able to tell the difference between this rare tumour and other health conditions.

These health conditions may present similarly to adamantinoma - in terms of symptoms and signs - but it is important the correct diagnosis is made to ensure the treatment provided is suitable. Conditions other than adamantinoma which can be the cause of particular symptoms are known as '**differential diagnoses**'.

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

- **OSTEOFIBROUS DYSPLASIA -** a benign (non-cancerous) childhood condition which regresses at the age of around 20 years old
- **FIBROUS DYSPLASIA -** a rare bone disorder which mostly occurs in adolescents and young adults
- **OSTEOMYELITIS -** the inflammation of the bone, or bone marrow, due to a bacterial infection
- **A NON-OSSIFYING FIBROMA -** a benign (non-cancerous) childhood tumour of the bone, which tends to occur in the shin bone – much like an adamantinoma

# TREATING ADAMANTINOMA

If the presence of adamantinoma 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.

It is known that there is little-to-no benefit of radiotherapy or chemotherapy in the treatment of adamantinoma. However, as chemotherapy reaches the whole body, it may be used if there is a spread of the tumour.

## SURGERY

The most effective treatment method for adamantinoma 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 adamantinoma 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 adamantinoma 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

Early diagnosis and adequate treatment, with wide surgical removal of the tumour, leads to an excellent outlook for patients with adamantinoma.

# 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 IS THE LEADING CHARITY DEDICATED TO FIGHTING PRIMARY BONE CANCER.

OUR MISSION IS TO SAVE LIVES AND IMPROVE OUTCOMES FOR PEOPLE AFFECTED BY PRIMARY BONE CANCER THROUGH RESEARCH, INFORMATION, AWARENESS AND SUPPORT.

**WE RECEIVE NO GOVERNMENTAL FUNDING, SO RELY ENTIRELY ON THE SUPPORT OF THE PUBLIC TO CONTINUE OUR LIFE SAVING WORK.**

# FOR INFORMATION AND SUPPORT CONTACT US:

**CALL 0113 258 5934**

**OR VISIT BCRT.ORG.UK**

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