RESEARCH
INFORMATION
AWARENESS
SUPPORT

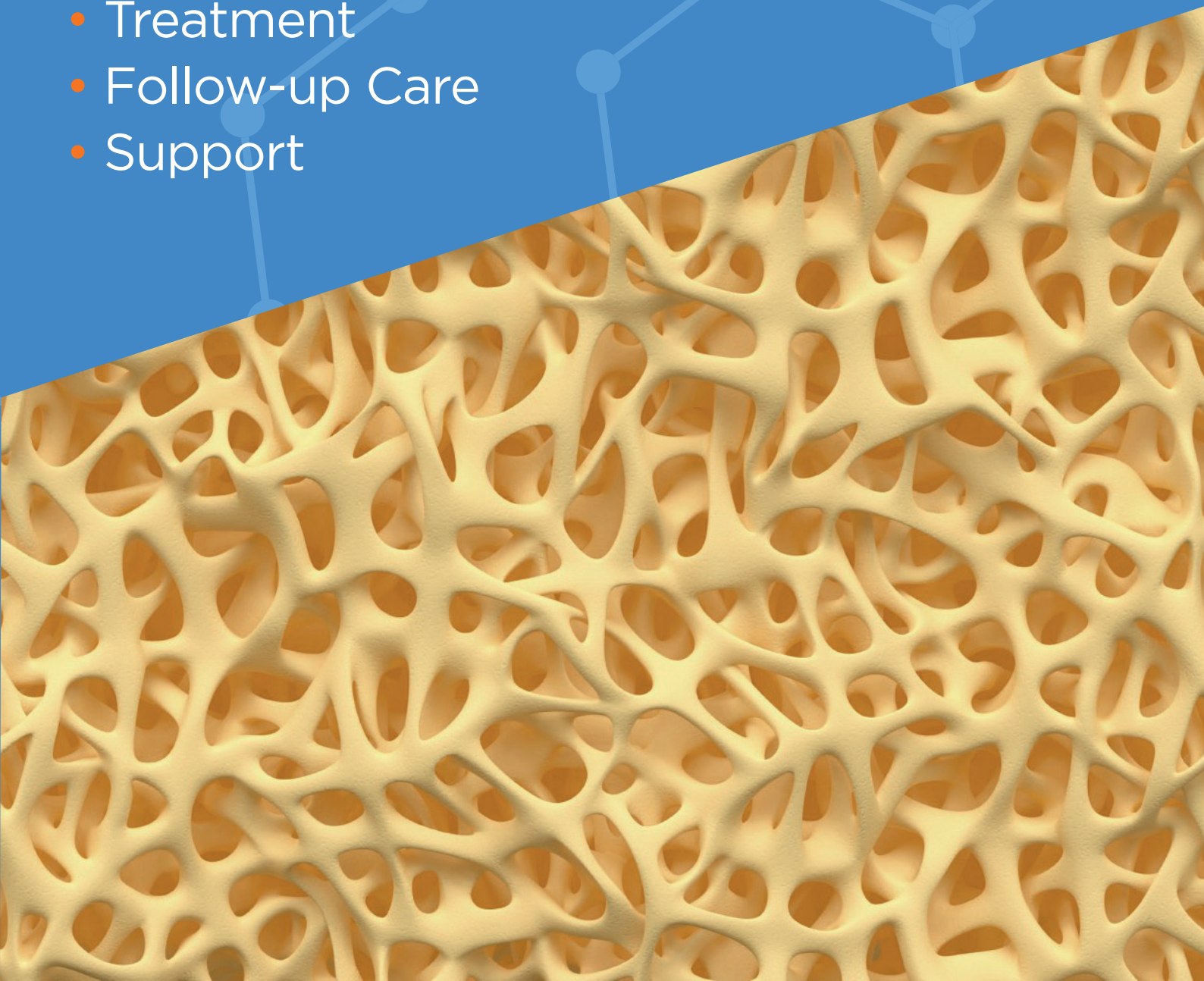
PRIMARY BONE CANCER

CHONDROSARCOMA

BONE CANCER RESEARCH TRUST
UNTIL THERE'S A CURE

Visit **bcrt.org.uk**
for more information

CONTENTS



Chondrosarcoma is the most common form of primary bone cancer affecting adults over the age of 30 years old. The most effective treatment strategy for this tumour type is surgery, which may in some cases be combined with chemotherapy or radiotherapy.

WHAT IS IT?

Chondrosarcoma is a rare cancer that most commonly forms in the bone. On very rare occasions, this tumour can also appear in the **soft-tissue** - which includes the ligaments, tendons, muscles and fat – where it is referred to as an '**extraskeletal chondrosarcoma**'.

This form of primary bone cancer is made up of tumour cells that produce **cartilage**, which is known as **chondroid** tissue. Cartilage is a rigid yet flexible connective tissue which is often found in the joints.

Chondrosarcoma can develop in any part of the skeleton, but the most common sites are the bones in the **legs**, the **upper arm bone**, the **pelvis**, the **shoulder blade** or the **rib cage**. In rare cases, chondrosarcomas can also develop in the spine or the skull.

WHO DOES IT AFFECT?

Chondrosarcoma is the most common form of primary bone cancer in adulthood and the second most common form of primary bone cancer affecting all age groups. It accounts for 25% of all primary bone cancer cases.

Chondrosarcoma can affect anyone, at any age, but is most commonly found in adults between the ages of 30 and 60 years of age and 80% of cases are in people aged 40 years or older.

In the UK alone, there are 190 new cases of chondrosarcoma diagnosed each year.

25%

OF ALL PRIMARY BONE CANCER CASES

WHAT ARE THE SYMPTOMS?

The symptoms of chondrosarcoma are general and can be similar to sports injuries, growing pains, or common conditions such as tendonitis or arthritis.

They may present for a long time before a diagnosis is made. Chondrosarcomas may also appear with no symptoms at all, and are detected when a patient suffers from a bone fracture or mild injury in 20-30% of cases. The most commonly reported symptoms of chondrosarcoma are:

BONE PAIN

which may be dull and can occur when a patient is at rest, often becoming worse at night

WALKING WITH A LIMP

RESTRICTED MOBILITY IN THE JOINTS NEAR THE AFFECTED BONE

A LUMP OR SWELLING

If the tumour is present on the spine, **neurological symptoms** such as muscle spasms, tingling sensations or muscle weakness may be observed

A BONE FRACTURE

These symptoms may vary in each patient and can present alone or in combination with one another.

TYPES OF CHONDROSARCOMA

By looking closely under a microscope at what kind of cells are making up the tumour, doctors can class each patient's tumour as one of **FOUR** main subtypes of chondrosarcoma. These are:

CONVENTIONAL CHONDROSARCOMA

Conventional chondrosarcomas make up around 85% of all chondrosarcoma cases and tend to occur in adults over the age of 50. Although they may affect any bone, they have a tendency to occur in the thigh bone, upper arm bone or the ribs.

DEDIFFERENTIATED CHONDROSARCOMA

Dedifferentiated chondrosarcomas make up around 10% of all chondrosarcoma cases and tend to be slightly more aggressive than conventional chondrosarcoma. Although they may affect any bone, they have a tendency to occur in the thigh bone or the pelvis in patients between 50 and 60 years of age.

MESENCHYMAL CHONDROSARCOMA

Mesenchymal chondrosarcomas make up less than 2% of all chondrosarcoma cases and are therefore extremely rare. This chondrosarcoma subtype tends to occur in teenagers and young adults and can often resemble Ewing sarcoma.

CLEAR CELL CHONDROSARCOMA

Clear cell chondrosarcomas make up less than 2% of all chondrosarcoma cases and are therefore extremely rare. This subtype is a low-grade tumour and tends to affect those aged between 30 and 40 years of age.

The classification of chondrosarcoma allows the best treatment plan to be implemented for each individual patient.

Chondrosarcoma may also be classified depending on their location in the bone. They may be classed as **CENTRAL CHONDROSARCOMAS**, located inside the bone cavity, or **SURFACE CHONDROSARCOMAS**, located on the outer surface of the bone.

CAUSES AND RISK FACTORS

There is no known cause of chondrosarcoma. However, there are possible causes and factors that have been identified which may increase an individual's risk of developing this tumour.

These risk factors include:

- **BEING OVER THE AGE OF 40 YEARS OLD**
- **HAVING A NON-CANCEROUS TUMOUR OF THE BONE -** such as chondroma or osteochondroma
- **UNDERGOING PREVIOUS RADIOTHERAPY**
- **HAVING UNDERLYING BONE ABNORMALITIES –** such as paget's disease, ollier's disease or maffucci syndrome
- **INHERITING CONDITIONS FROM YOUR PARENTS –** such as multiple hereditary exostoses or Wilms tumour
- **INHERTING DAMAGED GENES FROM YOUR PARENTS –** such as genes IDH1 and IDH2, which are frequently damaged in chondrosarcoma

DIAGNOSING CHONDROSARCOMA

Further tests to confirm a chondrosarcoma 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 doctor, where a **clinical examination** and an **X-ray** will be carried out. There is no clear sign that doctors can easily look for to make a diagnosis of chondrosarcoma. A CT scan and MRI scan provide crucial information on the exact location of the tumour, the stage of the tumour and the presence of a chondrosarcoma spreading elsewhere in the body.

Taking a biopsy of the bone is very useful when diagnosing chondrosarcoma. 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 chondrosarcoma.

AN ALTERNATIVE DIAGNOSIS?

When diagnosing chondrosarcoma, it is important to distinguish this tumour from other health conditions, or more common occurrences such as 'growing pains' or a sports injury. There may be health conditions that present similarly to chondrosarcoma - in terms of symptoms and signs - but it is important the correct diagnosis is made to ensure the treatment provided is suitable. Distinguishing a disease from a similarly presenting condition or disease is known as **'differential diagnosis'**.

If the diagnostic tests show that the patient does not have chondrosarcoma, there are a number of other conditions that may be presenting, including:

- **ENCHONDROMA -** a benign (non-cancerous) tumour of the cartilage
- **FIBROUS DYSPLASIA -** a bone disease in which normal bone is replaced by weak bone-like tissue.
- **OSTEOMYELITIS –** an infection of the bone

TREATING CHONDROSARCOMA

If the presence of chondrosarcoma 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.

Although each patient will receive their own personal treatment plan, chondrosarcoma tends to be treated using surgery. Radiotherapy, proton therapy or chemotherapy may also be used in those tumours that are more advanced.

SURGERY

The aim of surgery is to remove the primary tumour to prevent its growth and spread to other areas of the body. The surgical removal of a chondrosarcoma requires '**wide-surgical margins**'; this means some healthy tissue is removed alongside the tumour to ensure all tumour cells are removed and there is a lower risk of the tumour returning at a later date.

The surgical procedure carried out to treat chondrosarcoma aims to completely remove the tumour while preserving as much of the normal function and cosmetic appearance of the limb as possible.

Common surgical procedures performed to treat chondrosarcoma are:

- **CURETTAGE SURGERY:** tumour cells are scraped out of the bone
- **CYROSURGERY:** following the removal of the tumour, remaining cancer cells are frozen out using liquid nitrogen
- **RESECTION:** the affected area of bone is removed
- **AN AUTOGRAFT/AUTOLOGOUS GRAFT:** 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
- **IRRADIATION/REIMPLANTATION:** damaged bone is removed and treated with radiation, destroying the cancer cells, before being put back into the body
- **AMPUTATION:** on very rare occasions, the removal of the whole limb is required due to the size or location of the tumour. If possible, a prosthetic limb will be made for the patient

CHEMOTHERAPY AND RADIOTHERAPY

Methods of chemotherapy and radiotherapy are used less frequently in the treatment of chondrosarcoma. However, they may be used alongside surgery to treat certain subtypes of chondrosarcoma, particularly those that are of a higher-grade or at risk of spreading elsewhere in the body - such as dedifferentiated chondrosarcoma or mesenchymal chondrosarcoma.

Radiotherapy may also be used when the surgical removal of the tumour is not possible due to the tumours location. It may also be used after surgery to ensure all cancer cells have been destroyed. Finally, chemotherapy and radiotherapy may be used to relieve the pain and discomfort that patients at an advanced stage of this tumour may experience – this is known as **palliative chemotherapy** or **palliative radiotherapy.**

PROTON BEAM THERAPY

Proton beam therapy is a new and advanced form of radiotherapy that allows a greater dose of radiation to be delivered with a lower effect to surrounding healthy tissues. This aims to provide a more effective treatment that is also safer. The proton beam radiation is delivered to a specific location in the body, using MRI or CT scans, to ensure the tumour is directly targeted and the areas around the tumour are not affected.

This treatment is currently being developed in the UK; however it can be accessed in Switzerland or the USA if this treatment is recommended.

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

Bone Cancer Research Trust
10 Feast Field, Horsforth, Leeds, LS18 4TJ
bcrt.org.uk | 0113 258 5934
Charitable Incorporated Organisation
(CIO) Number - 1159590
@BCRT /BoneCancerResearchTrust

Certified Member