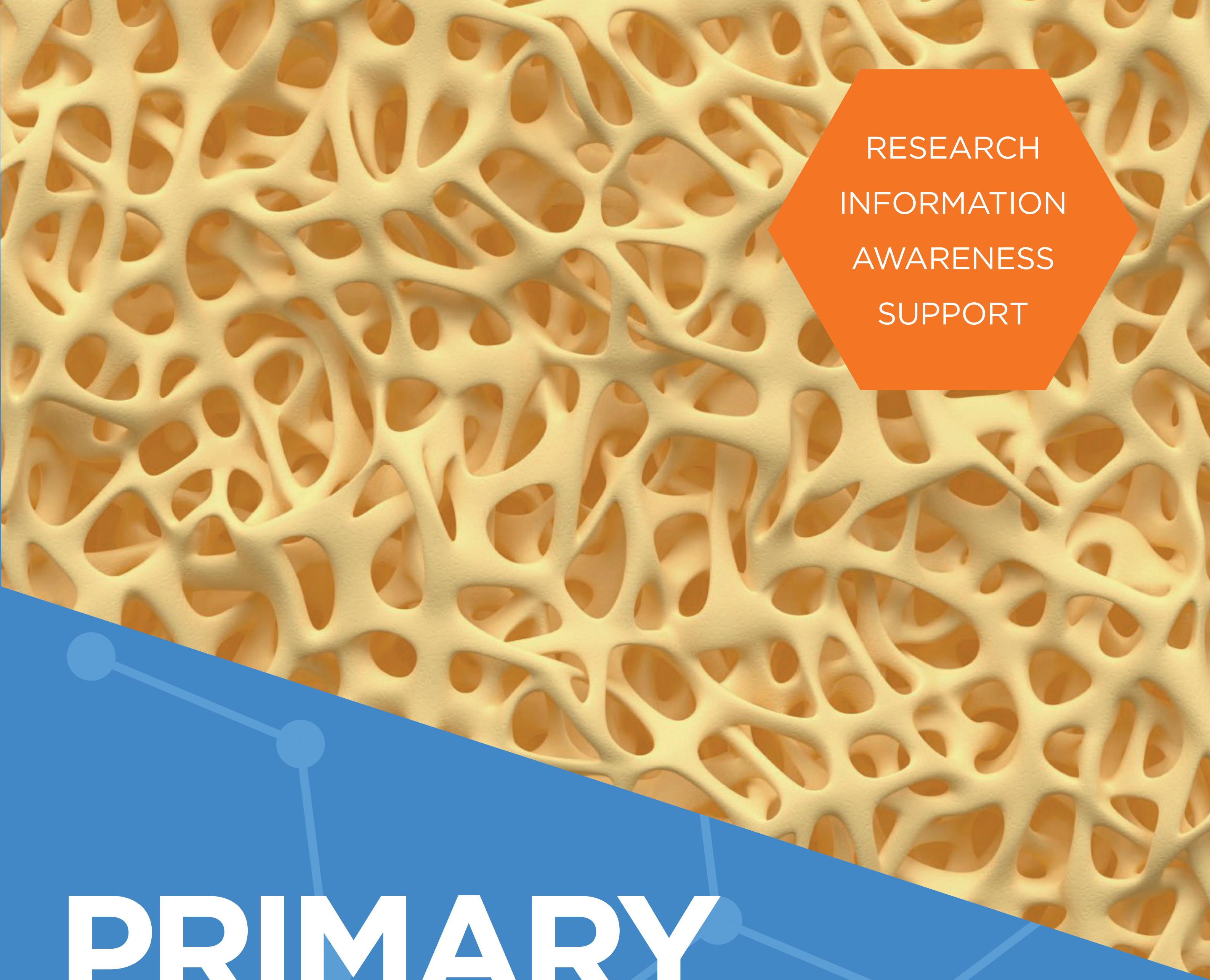

RESEARCH
INFORMATION
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PRIMARY BONE CANCER

AN OVERVIEW OF THE DISEASE, DIAGNOSIS AND TREATMENT

BONE CANCER RESEARCH TRUST
UNTIL THERE'S A CURE

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

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Primary bone cancer is the name given to a cancer that develops in a cell within the bone. This form of cancer, which may also be referred to as a bone sarcoma, is rare and makes up just 0.2% of all cancers diagnosed in the UK.

WHAT IS PRIMARY BONE CANCER?

Cancer develops when the body's cells behave in an abnormal way, growing and dividing uncontrollably to form a mass (or lump) of cells known as a tumour. The site at which a tumour forms is known as the 'primary tumour site'.

PRIMARY BONE CANCER IS THE NAME GIVEN TO A CANCER THAT DEVELOPS IN A CELL FOUND IN THE BONE

Primary bone cancer can develop in any bone in the body, though over a third of cases are found in the long bones of the lower body - such as the thigh bone (known as the femur) or the shin bone (known as the tibia).

If a cancer were to spread from its primary tumour site and form a tumour in another area of the body, this would be referred to as a 'secondary tumour site' or a 'metastatic site'. The bone is a common secondary tumour site and this is referred to as 'secondary bone cancer'.

WHO DOES IT AFFECT?

When combining known primary bone cancer incidences worldwide, we have estimated that a diagnosis of primary bone cancer is made **every 10 minutes** somewhere in the world. In the UK and Ireland alone, an average of 12 patients are diagnosed every week. Primary bone cancer is unusual in that it affects a wide range of age groups. It can affect children, adolescents, young adults and can also occur in older people; whereas the majority of cancers occur mainly in older adults. The most common forms of primary bone cancer are known to affect certain age groups:

- **Osteosarcoma and Ewing sarcoma most commonly affect children and young adults aged 10-25 years of age.**
- **Chondrosarcoma and chordoma most commonly affect adults over the age of 40 years old.**

EVERY 10 MINUTES

SOMEWHERE A CHILD, TEENAGER OR ADULT IS DIAGNOSED WITH PRIMARY BONE CANCER

Although mostly affecting those aged 10-25 years of age, osteosarcoma is also known to have a small incidence peak in people over the age of 55 years old.

WHAT ARE THE SYMPTOMS?

Symptoms of primary bone cancer can be very general and often appear similar to sports injuries, growing pains or many other common conditions such as tendonitis or arthritis. They may also vary depending on the location and the size of the tumour.

The most commonly reported symptoms of primary bone cancer are:

BONE PAIN

WHICH CAN BE CONSTANT OR MAY COME AND GO

PAINKILLERS MAY NOT HELP AND THE PAIN CAN BECOME WORSE AT NIGHT

TENDERNESS OF THE AREA

A LUMP OR SWELLING MAY BE SEEN OR FELT

AN UNEXPLAINED LIMP

A BONE FRACTURE

PROBLEMS WITH MOBILITY

SUCH AS STIFF JOINTS OR REDUCED MOVEMENT

BRUISING EASILY

TIREDNESS, FEVER, WEIGHT LOSS OR LOSS OF MUSCLE TONE MAY ALSO BE EXPERIENCED

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

TYPES OF PRIMARY BONE CANCER

By looking closely at what kind of cells are making up the tumour, doctors can class each primary bone cancer as a specific type.

85% of all primary bone cancers diagnosed are of the 3 most common types, which are:

OSTEOSARCOMA

Osteosarcoma is the most common form of primary bone cancer affecting children and young adults. It forms when a bone cell (known as an osteoblast) becomes abnormal and grows in an uncontrolled manner.

EWING SARCOMA

Ewing sarcoma is the second most commonly diagnosed form of primary bone cancer in children and young adults. It can develop anywhere in the body, though is most commonly diagnosed in the legs or pelvis.

EWING SARCOMA
IS THE
2ND
MOST
COMMONLY DIAGNOSED
FORM OF PRIMARY BONE CANCER
IN CHILDREN AND YOUNG ADULTS

CHONDROSARCOMA

Chondrosarcoma is the most common form of primary bone cancer occurring in adulthood, and accounts for 25% of all primary bone cancer cases. Chondrosarcoma develops from cells which produce cartilage and can occur anywhere in the body.

Additionally, there are 5 less common tumour forms that may arise in the bone, these are:

CHORDOMA

Chordoma accounts for less than 5% of all primary bone cancer cases and is most commonly found in the spine or the skull.

SPINDLE CELL SARCOMA OF THE BONE

Spindle cell sarcoma is a soft-tissue tumour which can also arise in the bone. This rare form of primary bone cancer tends to follow the diagnosis and treatment methods of osteosarcoma.

ADAMANTINOMA

Adamantinoma is very rare, accounting for less than 1% of all primary bone cancer cases, and is most frequently found in the shin bone.

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

ANGIOSARCOMA OF THE BONE

Angiosarcoma is a tumour developing 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.

GIANT CELL TUMOUR OF THE BONE

Giant cell tumours of the bone are non-cancerous tumours that most commonly arise around the knee. They are most effectively treated by surgically removing the tumour.

For further information on any of these primary bone cancer types please visit our website at **bcrt.org.uk**

DIAGNOSING PRIMARY BONE CANCER

Further tests to confirm a primary bone cancer diagnosis include:

- **AN X-RAY**
- **A CT SCAN**
- **AN MRI SCAN**
- **A PET SCAN**
- **A BONE SCAN**
- **A BIOPSY OF THE BONE**
- **BLOOD TESTS**

The first step in diagnosing any primary bone cancer is a trip to the GP. The GP will follow a specific set of guidelines when diagnosing a suspected primary bone cancer and will request an **X-ray** for the patient. It is very common for patients to be referred to a bone cancer specialist for a second opinion and confirmation of the diagnosis following an X-ray.

Further scans, such as a **CT scan**, an **MRI scan**, a **PET scan** and a **bone scan** are likely to be carried out once referred to a specialist in order to determine the location, size and extent of the suspected tumour.

CT SCAN

This scan takes a number of X-ray pictures from different angles to form a 3D image of the area of the body being examined. It should take around 10-20 minutes to complete and during this time the patient will be required to lie flat on a bed as it moves through the scanner.

MRI SCAN

MRI stands for ‘magnetic resonance imaging’. This scan is similar to a CT scan, but magnetism and radio-waves are used instead of X-rays to build up a detailed 3D image of the tumour. The patient will be required to lie still on a bed during the MRI scan, which provides a picture of where the tumour is in the bone and can help doctors decide on the right treatment plan for the patient.

PET SCAN

PET stands for ‘positron emission tomography’. These scans examine the whole body and can therefore take around an hour to complete. Before the scan, the patient will receive an injection of a harmless substance known as a radiotracer, which is taken into cells that are active. As cancer cells grow and divide much more rapidly than healthy cells, they take up the radiotracer much more and can be easily located during the PET scan to create a 3D image showing the number of cancer cells in a certain area.

BONE SCAN

Bone scans look for abnormalities in the bones. During the diagnosis of a suspected primary bone cancer, patients will often have a full body bone scan. Before having the scan, the patient will receive an injection containing a tiny amount of a harmless substance known as a radionuclide. This radionuclide is taken up by the bones over a few hours and can be detected by a gamma camera. The radionuclide will collect in larger amounts in cancerous areas allowing the location of the tumour to be determined.

These scans provide information on the size, location and extent of the tumour. However, in order to confirm the diagnosis and determine the specific type and grade of the tumour, a biopsy is required. A biopsy is a specialist procedure that removes a small sample of the tumour so it can be examined under a microscope. The small sample may be taken by a ‘needle biopsy’, to draw out a small amount of the tumour tissue using a needle, or by an ‘open biopsy’, to remove a small amount of tumour tissue during a small, minor, operation.

Results from a biopsy can take up to two weeks to analyse but they enable doctors to confirm the presence, and specific type, of primary bone cancer. For further information on the diagnosis of primary bone cancer, or any of the diagnostic tests discussed, please visit our website at bcrt.org.uk.

BONE CANCER CENTRES

Patients are referred to their nearest 'Bone Cancer Centre' by their GP for further diagnostic tests, and if required, treatment.

The Bone Cancer Centres have many experienced and specialised staff making up a multi-disciplinary team (MDT) of surgeons, oncologists, clinical nurse specialists and many more professionals.

If the presence of primary bone cancer is confirmed, the specialist medical team at the Bone Cancer Centre will design the best possible treatment plan for the individual patient.

For patients whose nearest Bone Cancer Centre is too far away, a 'shared care' arrangement with a closer hospital may be set up. This means that the specialist bone cancer centre recommends a treatment plan for the individual patient, which is then carried out at a hospital closer to the patient's home.

- **BONE CANCER SURGEONS -**
  who carry out the surgery required for the treatment of primary bone cancer and are specialised in this area of surgery
- **ONCOLOGISTS -**
  who are doctors specialising in looking after people with cancer
- **RADIOLOGISTS -**
  who are doctors specialising in looking at the images produced by X-rays, and other diagnostic scans, to help diagnose and assess cancer
- **CLINICAL NURSE SPECIALIST OR ADVANCED NURSE PRACTITIONER –**
  who are nurses specialising in a certain area, offering patients support and advice. Every person diagnosed with primary bone cancer should have a 'named nurse' who the patient has direct contact with and can talk to if they have any questions or concerns.
- **SOCIAL WORKERS AND PSYCHOLOGISTS –**
  who will help with patients' emotional, social and educational needs
- **DIETICIANS –**
  who will help ensure a patient is receiving all the nutrients and dietary needs they require during treatment and recovery

TREATING PRIMARY BONE CANCER

Specialists from many different areas will decide on the best treatment plan for each individual patient as a multi-disciplinary team (MDT). The main treatments a primary bone cancer patient will undergo are surgery, chemotherapy and radiotherapy.

Each patient, and tumour type, will require different treatments tailored to the individual. This section aims to provide a brief overview of the main treatment methods used for primary bone cancer patients. For more information on the treatment methods for specific tumour types please refer to our website at bcrt.org.uk.

SURGERY

Surgery is carried out with the aim to completely remove the primary tumour, so that it can no longer grow or spread, while having as little effect to the body as possible. In the majority of cases, limb-salvaging surgery will be carried out in order to achieve this.

COMMON LIMB-SALVAGING SURGERY PROCEDURES ARE:

- **SURGICAL RESECTION:** if possible, the tumour will be removed with little-to-no effect to the body's function and no need to reconstruct the area of affected bone
- **AN AUTOGRAFT (OR AUTOLOGOUS BONE GRAFT):** the affected area of bone is removed and reconstructed using the patients' own healthy bone 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 surgically removed
- **A METALLIC REPLACEMENT/IMPLANT:** once the tumour is removed the area of damaged bone is replaced with a metal implant known as a prosthesis. The prosthesis is usually measured and made specifically for each patient.
- **IRRADIATION/RE-IMPLANTATION:** the area of affected bone will be removed, treated with radiation to destroy the tumour cells and then put safely back into the patient.

These types of procedures are known as limb-salvaging surgery, of which 90-95% of patients will undergo. While this type of surgery is effective for most patients, less than 10% of patients may need to have the limb removed in order to remove the whole tumour and dramatically reduce the risk of the cancer returning at a later date. This is called an amputation. Once the area is healed following an amputation, a prosthetic (artificial) limb can be made specifically for the patient.

Patients may require additional surgery at a later date in order to rebuild the affected limb following the tumour's removal and, particularly if the patient is young and still growing, to replace or expand a prosthesis as the patient grows.

Following bone or joint surgery, patients will require a lot of support and physiotherapy to get going again. For information regarding the support a patient may receive after surgery please see our website (bcrt.org.uk) or ask your medical team for more advice.

CHEMOTHERAPY

Chemotherapy, often referred to as 'chemo', is the treatment of cancer using drugs. These drugs work by stopping the growth and division of cancer cells, or by damaging the cancer cells' DNA which causes these cells to die.

In order to be effective, chemotherapy drugs must take advantage of the difference between cancer cells and normal cells. Cancer cells divide much more rapidly than normal cells and therefore chemotherapy drugs target this rapid cell division process. Different chemotherapy drugs target slightly different parts of the cells' division machinery, and so chemotherapy is often used as a mix of multiple drugs - referred to as 'combination therapy' - in order to target the tumour more effectively.

Most healthy cells do not divide very quickly, however some types do; such as the cells producing hair, the reproductive cells, bone marrow cells, and the cells lining the digestive system. This means chemotherapy drugs can also affect these healthy, quick-dividing, cells and this is what causes the side-effects that some patients suffer from during chemotherapy treatment. Side-effects can be often be unpleasant and include:

- **NAUSEA**
- **DIARRHOEA**
- **HAIR LOSS**
- **MOUTH SORES**
- **AN UNUSUAL TASTE IN THE MOUTH**
- **TIREDNESS**
- **A LOW NUMBER OF WHITE BLOOD CELLS**
- **WEIGHT LOSS**

Each patient is different and so may not experience the same side effects, or may not get them as severely, as others may.

FOR INFORMATION ON HOW TO DEAL WITH THESE SIDE EFFECTS PLEASE VISIT OUR WEBSITE AT BCRT.ORG.UK, OR ASK YOUR MEDICAL TEAM FOR MORE ADVICE.

Chemotherapy is given to patients in '**cycles**', which involve treatment and rest periods. Patients tend to have the chemotherapy drug, or combination of drugs, over 3-4 days before having a rest period of around 2-3 weeks to allow the body to recover. This is classed as one cycle, and will be repeated over a set amount of time. Primary bone cancer patients tend to have up to 12 weeks of chemotherapy before surgery and a possible further 30 weeks afterwards; though the specific duration and drugs used will differ between each patient and tumour type.

Chemotherapy is administered through the blood stream in order to reach different areas of the body and target all the cancerous cells. The most common ways of taking chemotherapy drugs include:

- **SWALLOWING A TABLET OR LIQUID MEDICINE**
- **BY AN INJECTION**
- **PLACING AN INTRAVENOUS (IV) TUBE INTO A VEIN**
- **BY A CENTRAL LINE OR PICC (PERIPHERALLY INSERTED CENTRAL CATHETER).** These are long flexible tubes which are put in while the patient is under general or local anaesthetic.This method is often used in patients having an intensive course of chemotherapy and the tubes stay in throughout treatment to reduce the number of needles and injections needed.

RADIOTHERAPY

Radiotherapy is used less frequently in the treatment of primary bone cancer in comparison to surgery or chemotherapy. However, it may be used following surgery for the treatment of Ewing sarcoma, chordoma and occasionally osteosarcoma.

Unlike chemotherapy, radiotherapy is a **'local therapy'** meaning it is designed to treat the tumour directly rather than reach the whole body. Radiotherapy uses a high-energy, focused radiation beam (usually an X-ray) to permanently damage the DNA inside of the tumour cells; this damage prevents the cells from dividing and causes them to die.

Radiotherapy can be used to shrink the tumour before surgery, remove any remaining tumour cells in the area after surgery or be used in patients who cannot undergo surgery due to the location of their tumour (for example a tumour in the pelvis or spine).

The benefit of radiotherapy must be balanced against the possible unpleasant side-effects patients may experience from this treatment. Side-effects will depend on the location the radiation was applied to, but most commonly include:

- **SORE SKIN OR A SKIN REACTION**
- **TIREDNESS**
- **A DRY MOUTH AND DIFFICULTY SWALLOWING**
- **NAUSEA**
- **LOSS OF APPETITE**
- **DIARRHOEA**

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. It may be best to talk to your medical team about these possible late-effects.

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:** psychologists will support patients through any feelings of anxiety, help to deal with patient concerns or overcome the psychological impact a cancer diagnosis or treatment may bring
- **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
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@BCRT /BoneCancerResearchTrust

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