CONTENTS

• What is it?
• Who does it affect?
• Symptoms
• Biology of Ewing Sarcoma
• Cause and Risk Factors
• Diagnosis
• Treatment
• Follow-up Care
• Support
Ewing Sarcoma is a form of primary bone cancer that most commonly affects children and young adults. This form of cancer belongs to a group of tumours known as the ‘Ewing Sarcoma Family of Tumours’ and can develop anywhere in the skeleton.

**WHAT IS IT?**

85% of the Ewing sarcoma family of tumours develop in the bones. The Ewing sarcoma family of tumours includes:

- **Ewing sarcoma** (a form of primary bone cancer)
- **Extraosseous Ewing sarcoma** (Ewing sarcoma of the soft tissue)
- **Askin Tumour** (Ewing sarcoma that starts in the chest wall)
- **Primitive Neuroectodermal Tumour** (Ewing sarcoma in which the cells look like nerve cells)

Ewing sarcoma can develop anywhere in the skeleton, but is most commonly found in the flat bones of the body, which include the bones in the **legs**, the **pelvis** and the bones in the **chest**.

**WHO DOES IT AFFECT?**

Following osteosarcoma, Ewing sarcoma is the second most commonly diagnosed form of primary bone cancer in children and young adults - accounting for 1.5% of all childhood cancers.

Although it can develop in anyone, at any age, Ewing sarcoma is rare in people over 30 years of age and occurs most frequently in individuals aged between 10 and 20 years old, who are experiencing periods of rapid bone growth.
WHAT ARE THE SYMPTOMS?

The symptoms of Ewing sarcoma may be mild at first and slowly progress, or they may suddenly appear. Symptoms are general and can be similar to sports injuries, growing pains, or common conditions such as tendonitis or arthritis.

The most commonly reported symptoms of Ewing sarcoma include:

- **Bone Pain** which may be constant or come and go, and may be worse at night
- **A Lump or Swelling**
- **A Bone Fracture**
- **Tiredness, Fever, Weight Loss and Breathlessness** may also be experienced
- **The Sensation of Pins and Needles**

These symptoms may vary in each patient and can present alone or in combination with one another.
We know that a large proportion of Ewing sarcoma patients share the same kind of gene damage - although we don’t know what causes this damage to occur.

Genes are made of a molecule called DNA. In cells, DNA is found in long strings called chromosomes. Each chromosome is made up of thousands of genes, one after another.

We know that in Ewing sarcoma there is a chromosomal translocation – this means that a part of a chromosome has broken off and stuck to the wrong chromosome. This puts genes in the wrong order and can mean that genes are switched on and off incorrectly.

Damage to these specific genes and the production of EWS-FLI1 causes the tumour cells to behave differently and grow abnormally - leading to development of cancer. The presence of EWS-FLI1 is used to help confirm the diagnosis of Ewing sarcoma.

IN 95% OF EWING SARCOMA PATIENTS, A CHROMOSOMAL TRANSLOCATION TAKES PLACE WHICH INCORRECTLY STICKS TWO GENES TOGETHER TO MAKE A ‘FUSION GENE’ KNOWN AS EWS-FLI1.
CAUSES AND RISK FACTORS

The cause of Ewing sarcoma is largely unknown. Therefore, there is no known way of preventing this cancer.

There is a known relationship between periods of rapid bone growth in adolescents and the development of primary bone cancers such as Ewing sarcoma and osteosarcoma.

There are no confirmed links between any environmental factors or inherited risk factors and the development of Ewing sarcoma. However, there are possible factors that have been identified which may increase an individual’s risk of developing Ewing sarcoma.

These risk factors include:

• BEING AGED BETWEEN 10 AND 20 YEARS OF AGE
• BEING CAUCASIAN
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 Ewing sarcoma. 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 Ewing sarcoma spreading elsewhere in the body.

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

Further tests to confirm an Ewing sarcoma diagnosis include:

• A CT SCAN
• AN MRI SCAN
• A BIOPSY OF THE BONE
• A BONE MARROW BIOPSY
• BLOOD TESTS
When diagnosing Ewing sarcoma, 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 Ewing sarcoma 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 Ewing sarcoma, there are a number of other conditions that may be presenting, including:

- **OSGOOD SCHLATTERS DISEASE** - which is found in physically active adolescent boys and girls, and is caused by stress on the tendons that connect to the knee-cap.

- **A SLIPPED EPIPHYSIS** - the growing section of bone, present at the end of the long bone, slips and moves on the bone which can cause bone pain.

- **EOSINOPHILIC GRANULOMA** - a benign (non-cancerous) bone tumour that is found mainly in children under the age of 10 years.

- **OSTEOMYELITIS** - an infection of the bone.

- **OSTEOSARCOMA** - another form of primary bone cancer.

- **CHONDROSARCOMA** - another form of primary bone cancer.
TREATING EWING SARCOMA

If the presence of Ewing sarcoma 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. Managing Ewing sarcoma involves treatment to the whole body using chemotherapy and treatment of the tumour site directly using surgery and radiotherapy.

CHEMOTHERAPY

NEO-ADJUVANT CHEMOTHERAPY

In most cases, chemotherapy is used before surgery to kill cancer cells and shrink the tumour to make surgery easier; this is known as ‘neo-adjuvant chemotherapy’.

In the UK, Ireland, and much of Europe, the standard chemotherapy treatment given before surgery is made up of four drugs: Vincristine, Ifosfamide, Doxorubicin and Etoposide (this is referred to as ‘VIDE’).

ADJUVANT CHEMOTHERAPY

Chemotherapy can also be used after surgery to destroy any remaining cancer cells in the location of the tumour, or in the rest of the body; this is known as ‘adjuvant chemotherapy’.

In the UK, Ireland, and much of Europe, the standard chemotherapy treatment given after surgery is made up of four drugs: Vincristine, Actinomycin D, Ifosfamide and Cyclophosphamide.

Types of chemotherapy drugs used to treat Ewing sarcoma are:

- **VINCristine**
- **IFOSfamide**
- **DOXORubcin**
- **ETopoSIDE**
- **ACTINOMYCIN D**
- **CYCLOPHOSPHAMIDE**
- **TEMOZOLAMIDE**
- **IRINOTECAN**
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 an Ewing sarcoma 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 an Ewing sarcoma is known as ‘**limb-salvaging surgery**’, which aims to completely remove the tumour while preserving as much of the normal function and cosmetic appearance of the limb as possible.

Common limb-salvaging surgical procedures performed are:

- **RESECTION:** the affected area of bone is removed
- **AN AUTOGRRAFT/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 require 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

RADIOThERAPY

Ewing sarcoma responds well to radiotherapy treatment. Radiotherapy is sometimes used instead of surgery in cases where surgical removal of the primary tumour is not possible - for example if the tumour is in the pelvis or spine.

Radiotherapy may also be given after surgery to ensure all tumour cells in the area are destroyed. The exact dose and length of radiotherapy treatment will also be decided by a specialist team of doctors, but it is usually given as a single dose each day (lasting a few minutes) for approximately **5 – 6 weeks**.

HIGH DOSE CHEMOTHERAPY WITH AUTOLOGOUS STEM CELL TRANSPLANT

Some Ewing sarcoma patients may be offered high-dose chemotherapy with an ‘autologous stem-cell transplant’. This allows the chemotherapy to have a higher impact on the cancer while replacing any damaged cells that this treatment method causes.

Bone marrow stem cells are immature cells that can differentiate into any blood cell type. These stem cells are collected using a specialised machine before high dose chemotherapy begins, in a process known as ‘**stem cell harvesting**’.

Once removed, the stem cells are stored and frozen until the high-dose chemotherapy is complete and the stem cells are then transplanted back to the patient using a drip. This process replaces the blood cells that are damaged or even destroyed during chemotherapy.
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:

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