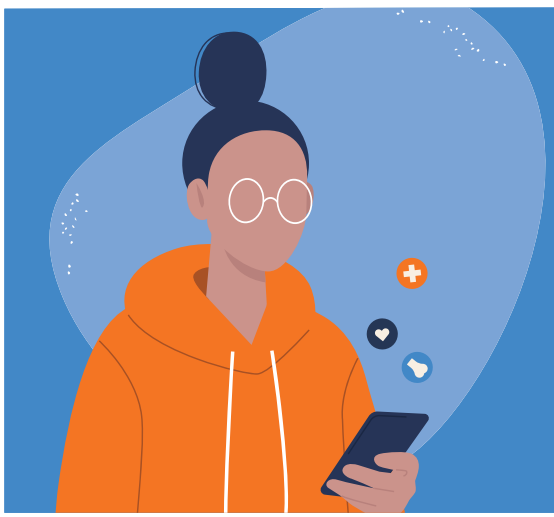

Key points about

EWING SARCOMA

V1.0: OCT 23

BONE CANCER RESEARCH TRUST

Trusted Information Creator
Patient Information Forum

1. Who this leaflet is for

This leaflet has been written for people aged over 16 years old to help them understand Ewing sarcoma, its diagnosis and treatment. Young people under 16 years old may also find it useful.

2. What Ewing sarcoma is

Ewing sarcoma is a rare cancer that can start in the bones or in soft tissue, like the muscles. It belongs to a group of tumours called 'undifferentiated small round cell sarcomas of the bone and soft tissue'. Ewing sarcoma more commonly starts in the bones. In particular, the pelvis, thigh bone (femur) and ribs.

3. Causes of Ewing sarcoma

A large number of Ewing sarcoma tumours share the same kind of gene damage, where 2 separate genes (DNA) combine (fuse) to become a 'fusion' gene. Most Ewing sarcomas are caused by fusions between the same 2 genes. Doctors do not yet know what causes fusion between certain genes, or how to stop it happening. We do know that:

- You cannot catch Ewing sarcoma from anyone else.
- There do not seem to be any environmental factors that cause it, such as radiation.
- It is not caused by something you have done.

4. Who Ewing sarcoma affects

Ewing sarcoma is the second most common type of primary bone cancer found in young people. It most commonly affects people between the ages of 10 to 24 years. However, it can also affect older adults and younger children.

Each year in the UK around 90 people are diagnosed with Ewing sarcoma.

5. Symptoms of Ewing sarcoma

The symptoms of Ewing sarcoma are general and can be similar to other conditions. They may be mild at first and slowly progress, or they may suddenly appear. The symptoms and number of symptoms vary for every person. Symptoms are:

- bone pain which might be worse at night, happens all the time or stops and starts, is not helped by painkillers and may get worse over time
- a lump or swelling, which may be seen or felt if the tumour is near the skin's surface (called a 'palpable mass')
- a broken bone caused by weakening of bone, due to a tumour, without having had a fall or accident (called a 'pathological fracture')
- problems moving, may develop a limp, stiff limbs or joints, unable to move as normal
- tenderness over the bone or joint
- a high temperature (fever), feeling tired or weary ('lethargy' or 'fatigue'), pain with tingling and numbness ('pins and needles'), weight loss and loss of appetite, feeling breathless

6. Diagnosing Ewing sarcoma

Doctors will carry out several different tests before a diagnosis of Ewing sarcoma can be confirmed. These tests may include:

- x-ray (usually carried out first)
- biopsy (where a small piece of tumour is collected and the cells looked at under a microscope)
- blood tests
- bone marrow biopsy
- CT scan, MRI scan, PET scan

X-rays and scans help doctors to see the size of the tumour and exactly where in the body it is. Scans also check if the cancer has spread to any other parts of the body (called 'staging'). Using the test results, the doctor will decide on a treatment plan.

7. Holistic Needs Assessment

This is where you have a chat with someone in your medical team who will ask you questions about your worries and concerns. It covers all parts of life. They will work with you to create a support plan. You may be offered an assessment at diagnosis, during treatment or after treatment.

8. Prognosis

When a person has an illness, doctors try to predict how it will affect that person. For example, how likely it is that the treatment will work, and the person will be cured. This is called a 'prognosis'.

Doctors will consider lots of things about you and the tumour when making a prognosis for Ewing sarcoma. Everyone is different, so doctors can never be sure how Ewing sarcoma will affect each person.

9. Treatments for Ewing sarcoma

Most people are usually diagnosed and usually have surgery at a bone cancer centre. Other treatments such as chemotherapy and radiotherapy may be given at a hospital closer to you. Treatments for Ewing sarcoma include:

Chemotherapy (chemo)

Chemotherapy (chemo) is a type of medicine, that travels around the whole body in the bloodstream. It is given in 'cycles', in most cases, both before and after surgery. The aim of chemo is to:

- shrink the tumour before surgery
- destroy any cancer cells that could have spread from the original (primary) site to other areas of the body

The standard chemo given to treat Ewing sarcoma before and after surgery is now internationally recognised as the VDC/IE drug combination from the Euro Ewings 2012 trial (EE2012). The medicines given are called Vincristine, Doxorubicin, Cyclophosphamide, Ifosfamide and Etoposide.

Surgery

After chemo and, or radiotherapy, you may have surgery. Its aim is to completely remove the primary tumour and keep the body working as normally as possible. For some people, it is very difficult to remove the tumour. For example, if the tumour is in the pelvis or the spine.

If the Ewing sarcoma is in a limb (arm or leg), your medical team will do their best to save the limb by doing 'limb-sparing surgery'. This is not always possible, and they may need to remove the limb. This is called an 'amputation'.

Your doctors and nurses will talk to you about your options. Visit our website to watch videos about amputation: **bcrt.org.uk/amputation**

Radiotherapy

Radiotherapy means treating the tumour with high doses of concentrated radiation. It is usually given as a single dose each day for about 5 to 6 weeks.

Radiotherapy may be used:

- before surgery to shrink the cancer
- after surgery to destroy any cancer cells that remain in the area
- instead of surgery if the tumour cannot be removed by surgery
- with surgery and chemo if the tumour cannot be fully removed or if full removal would limit how well the body works

Some people with Ewing sarcoma may be eligible for Proton Beam Therapy (PBT). This is a type of focused radiotherapy that delivers a radiation dose directly to the tumour.

10. Support after treatment

The end of your treatment will come as a welcome relief, but it can also be a time of worry and anxiety. You may like to join a support group or contact other people with primary bone cancer through blogs or on social media.

The Bone Cancer Research Trust's Support and Information Team can connect you with others who have experience of primary bone cancer. You can also find patient stories on our website at: **bcrt.org.uk/patientstories**

There are lots of other organisations who can support you too. They are listed on our website at: **bcrt.org.uk/usefulorganisations**

11. Follow-up care

Your doctors will want to monitor you for a long time after treatment (called 'follow-up care'). Outpatient hospital visits will be needed, the frequency of these will depend on each person.

Doctors will check your general health, talk about any concerns and do tests to:

- check if the cancer has come back (called 'relapse' or 'recurrence')
- check for any 'late effects' from the cancer treatment

You may also have follow-up care with a surgical team. In-between visits, it is important you get in touch with your key worker or doctor straight away if you have any problems.

12. If Ewing sarcoma comes back

Sadly, Ewing sarcoma can come back in some people. If the cancer does return, you may be given a different mix of chemo medicines. The treatment may be more aggressive than before. You may need more surgery and maybe radiotherapy, or be invited to take part in a clinical trial. Treatments will depend on each person. The doctors and nurses will talk you through the treatment options in detail.

13. Advanced Ewing sarcoma

Your doctor may have told you that the cancer can no longer be cured, and the focus now is to make you comfortable and give you as much time as possible with your loved ones. They may talk to you about palliative care, ways to ensure your pain is managed and hospice care. For more information on this, contact our Support and Information Team.

Also available:

- Your guide to Ewing sarcoma (booklet). Contact us to request your free copy.
- Our website. Visit: **bcrt.org.uk/ewingsarcoma**

For information about the references used to create this leaflet, or if you have any questions or feedback, contact our Support and Information Service:

Visit: bcrt.org.uk/support
Call: 0800 111 4855
Email: support@bcrt.org.uk

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Visit **bcrt.org.uk/ourhealthinformation** to find out how we produce our information resources and to read our disclaimer.

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