CONDITION

Ewing sarcoma

Ewing sarcoma is a cancer that usually starts in bone and mostly affects children, teenagers, and young adults.

WHAT IT IS
Cancer of bone or nearby soft tissue

BODY SYSTEM
Bones and soft tissues

MAIN SYMPTOMS
Persistent pain, swelling, tenderness, unexplained fracture

WHAT CAUSES IT
Acquired DNA change, not inherited

HOW IT IS DIAGNOSED
Biopsy with pathology and molecular testing

MAIN TREATMENT
Chemotherapy with surgery and/or radiation therapy

CAN IT BE CURED?
Sometimes - more likely when localized

IS IT AN EMERGENCY?
Sometimes - suspected fracture or serious breathing difficulty

LONG-TERM OUTLOOK
Localized cancer often curable; spread or relapse worsens outlook

Overview

Ewing sarcoma is a rare cancer that most often begins in a bone. It can also start in the soft tissues around bones, such as muscle or connective tissue.

Common starting points include the pelvis, ribs, chest wall, and long bones of the arms or legs. Ewing sarcoma mainly affects children, teenagers, and young adults, but it can occur at any age.

The cancer develops when a cell acquires a specific change in its DNA. DNA contains the instructions that control how cells behave. In Ewing sarcoma, parts of two genes usually become joined, creating an abnormal "fusion gene" that encourages uncontrolled cell growth.

Ewing sarcoma can grow and spread, so timely specialist assessment matters. Treatment

usually combines chemotherapy with surgery, radiation therapy, or both. The possibility of cure is highest when the cancer remains in its original area, known as localized cancer.

Symptoms

Pain is the most common symptom. It may come and go at first but often becomes more persistent. Pain can be worse during activity or at night, and it may not improve as expected after a minor injury.

Other possible symptoms include:

Symptoms depend partly on where the cancer is growing. A tumour in the chest wall may cause chest discomfort, coughing, or breathing problems. A tumour near the spine may press on nerves, causing weakness, numbness, or changes in bladder or bowel control.

These symptoms have many more common causes, including sports injuries, infections, and non-cancerous lumps. Symptoms alone cannot confirm Ewing sarcoma. Pain or swelling that persists, worsens, or has no clear explanation should be medically assessed.

- Swelling, tenderness, or a noticeable lump
- Reduced movement, stiffness, or a limp
- A bone that breaks more easily than expected
- Tiredness, fever, reduced appetite, or unplanned weight loss

Causes and risk factors

Ewing sarcoma is caused by DNA changes that develop within an affected cell. In most cases, a gene called EWSR1 becomes fused to another gene, most often FLI1 . This change is found only in the cancer cells.

The DNA change is not usually inherited from a parent, and people with Ewing sarcoma do not generally pass it to their children. It is not known to result from diet, exercise, an injury, or something a person or their family did.

Age is the clearest known pattern. The condition is most often diagnosed during adolescence or early adulthood, although younger children and older adults can also develop it. There is no established lifestyle-related risk factor that can reliably predict who will be affected.

Diagnosis

Diagnosis usually begins with imaging. An X-ray may show changes in the affected bone. MRI provides more detail about the tumour and nearby muscles, nerves, blood vessels, and joints.

A biopsy is needed to confirm Ewing sarcoma. During a biopsy, a specialist removes a small sample of the tumour. A pathologist, who is a doctor trained to identify conditions by examining cells and tissues, studies the sample under a microscope.

Laboratory testing looks for the gene fusion associated with Ewing sarcoma. This helps distinguish it from other cancers that can look similar under a microscope.

Whenever possible, the biopsy should be planned by the sarcoma team that would perform any later surgery. The position of the biopsy path can affect how the tumour is removed.

If Ewing sarcoma is confirmed, further tests show its size and whether it has spread. These may include:

The lungs, other bones, and bone marrow are the most important areas checked for spread. Bone marrow testing may be used in selected situations. The results allow the team to classify the cancer as localized or metastatic, meaning it has spread to another part of the body.

- MRI of the original area
- CT of the chest to examine the lungs
- PET-CT or a bone scan to examine other bones and tissues
- Blood tests to assess general health and organ function

Treatment and management

Treatment should be coordinated by a specialist sarcoma team. For children and teenagers, this is usually a paediatric cancer team. Care commonly involves cancer specialists, surgeons, radiologists, pathologists, nurses, physiotherapists, and other support professionals.

Chemotherapy is a central part of treatment because Ewing sarcoma cells may travel beyond the original tumour even when scans do not show spread. Several chemotherapy medicines are given in planned combinations over many months. Some treatment is usually given before surgery or radiation, followed by more chemotherapy afterward.

Treatment of the original tumour is called local control. It may involve:

When the cancer has spread, treatment usually still includes chemotherapy and treatment directed at the original tumour. Surgery or radiation may also be considered for areas of spread. The exact approach is based on where the cancer is, how it responds to chemotherapy,

and the person's overall health.

Treatment also includes medicines and practical support for nausea, pain, infection risk, nutrition, mobility, and emotional health. Chemotherapy and radiation may affect fertility, so fertility preservation should be discussed before treatment when time and circumstances allow.

A clinical trial may provide access to a treatment strategy being studied. Joining one is voluntary.

- Surgery: The surgeon removes the tumour with a border of healthy tissue when this can be done safely. Limb-sparing surgery is often possible. Amputation is sometimes needed when the tumour cannot otherwise be removed with an acceptable result.
- Radiation therapy: High-energy radiation damages cancer cells. It may be used when surgery is not possible, when removing the whole tumour would cause major harm, or when cancer cells may remain after surgery.
- Surgery and radiation: Some tumours require both approaches.

Complications

Ewing sarcoma may weaken a bone, leading to a fracture. Tumours can also affect nearby joints, nerves, blood vessels, or organs. Cancer that spreads most often reaches the lungs, other bones, or bone marrow.

Treatment can cause short-term effects such as tiredness, hair loss, nausea, mouth soreness, and low blood cell counts. Low white blood cell counts can make serious infections more likely.

Possible long-term effects depend on the medicines used, the radiation area, surgery, and age during treatment. They can include reduced joint movement, differences in bone growth, fertility problems, heart or kidney effects, and a small risk of another cancer later. Rehabilitation and long-term follow-up are used to identify and manage these problems.

Ewing sarcoma can return after treatment. A recurrence may be near the original site or in another area, commonly the lungs or bones. Recurrent cancer is more difficult to cure, but further treatment may control it or, in some circumstances, lead to long-term remission.

Prevention

There is no known way to prevent Ewing sarcoma. Its main DNA change is acquired by an individual cell and is not linked to a known avoidable behaviour or exposure.

Screening people without symptoms is not recommended because there is no established screening test for Ewing sarcoma. Families should not blame an injury, activity, or lifestyle choice for causing it.

Living with it and what to expect

Initial treatment usually lasts many months and can interrupt school, work, sports, and family routines. Hospital visits are frequent, and some chemotherapy may require hospital stays. Energy and appetite can change during each treatment cycle.

Physical therapy may help maintain movement and strength. After surgery, recovery depends on the bone involved and the type of reconstruction. Some people use a brace, crutches, or another mobility aid temporarily or long term.

After treatment, follow-up continues for years. Visits usually include examination and imaging of the original area and lungs. Appointments are more frequent at first and become less frequent over time. Follow-up also checks growth, mobility, heart and kidney health, fertility, emotional well-being, and other possible late effects.

Many people treated for localized Ewing sarcoma return to education, work, relationships, and regular activities. Some continue to have pain, reduced movement, fatigue, or concerns about recurrence. The course is different for each person and is strongly affected by whether the cancer had spread, its location, and how it responded to treatment.

If the cancer returns or cannot be cured, treatment may still shrink it, slow its growth, and relieve symptoms. Palliative care can be provided alongside cancer treatment to support comfort and quality of life; it is not limited to the final stage of life.

When to get medical help

Arrange a medical assessment for bone pain, swelling, or a lump that persists, worsens, wakes you at night, or has no clear explanation. A limp or reduced use of an arm or leg also deserves assessment, particularly when it does not improve.

If you are receiving chemotherapy, contact your cancer team immediately about a fever, shaking chills, or feeling suddenly unwell. Infections can become serious while white blood cell counts are low. Follow the team's specific instructions rather than waiting for a routine appointment.

Seek urgent or emergency care for:

Routine questions about pain control, treatment effects, rehabilitation, or follow-up scans can usually be discussed with the cancer team during normal contact hours unless symptoms are rapidly worsening.

- Sudden severe bone pain, deformity, or inability to use a limb
- Serious trouble breathing, collapse, or new confusion
- New leg weakness, numbness around the groin, or loss of bladder control
- Bleeding that does not stop

Common questions

Is Ewing sarcoma a type of bone cancer?

Usually. Most Ewing sarcomas begin in bone, but some start in nearby soft tissue. These tumours share the same characteristic cell and genetic features and are treated in similar ways.

Can Ewing sarcoma be cured?

Sometimes. Cure is more likely when the cancer is localized and responds well to chemotherapy. Cancer that has spread, especially to several bones or bone marrow, or that returns after treatment is harder to cure.

Is Ewing sarcoma hereditary?

It is not usually hereditary. The characteristic gene fusion develops in the cancer cell and is generally not present throughout the rest of the body. Most families do not need screening for Ewing sarcoma.

Does a painful lump mean I have Ewing sarcoma?

No. Pain and lumps are much more often caused by injuries, infections, cysts, or other non-cancerous conditions. Persistent or unexplained symptoms still need assessment because imaging and, when appropriate, a biopsy are required to identify the cause.

How quickly does Ewing sarcoma grow?

Ewing sarcoma can grow and spread relatively quickly, but the pace is not the same for everyone. Symptoms cannot show how far it has spread. Imaging and biopsy results provide that information.

Will surgery mean losing an arm or leg?

Not usually, because limb-sparing surgery is often possible. The options depend on the tumour's size and location and its relationship to nerves, blood vessels, and joints. Occasionally,

amputation provides the safest or most functional option.

Sources

- National Cancer Institute. Ewing Sarcoma Treatment-Health Professional Version . PDQ Cancer Information Summary.
- National Cancer Institute. Ewing Sarcoma Treatment-Patient Version . PDQ Cancer Information Summary.
- Grünewald TGP, Cidre-Aranaz F, Surdez D, et al. "Ewing sarcoma." Nature Reviews Disease Primers . 2018;4:5. doi:10.1038/s41572-018-0003-x.
- Strauss SJ, Frezza AM, Abecassis N, et al. "Bone sarcomas: ESMO-EURACAN-GENTURIS-ERN PaedCan Clinical Practice Guideline for diagnosis, treatment and follow-up." Annals of Oncology . 2021;32(12):1520-1536.
- American Cancer Society. Ewing Tumor . Diagnosis, treatment, and survivorship information.