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## About Ewing Tumors

### Overview and Types

If you or your child have just been diagnosed with a Ewing tumor or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- [What Is the Ewing Family of Tumors?](#)

### Research and Statistics

See the latest estimates for new cases of Ewing tumors in the US and what research is currently being done.

- [Key Statistics for Ewing Tumors](#)
  - [What's New in Ewing Tumor Research and Treatment?](#)
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## What Is the Ewing Family of Tumors?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see [What Is Cancer?](#)<sup>1</sup> For information about the differences between childhood cancers and adult cancers, see [Cancer in Children](#)<sup>2</sup>.

The Ewing family of tumors is a group of cancers that start in the bones or nearby soft

tissues that share some common features. These tumors can develop at any age, but they are most common in the early teen years.

The main types of Ewing tumors are:

- **Ewing sarcoma of bone:** Ewing sarcoma that starts in a bone is the most common tumor in this family. This type of tumor was first described by Dr. James Ewing in 1921, who found it was different from the more common bone tumor, [osteosarcoma](#)<sup>3</sup>. Seen under a microscope, its cells looked different from osteosarcoma cells. It was also more likely to respond to radiation therapy.
- **Extrasosseous Ewing tumor (EOE):** Extrasosseous Ewing tumors start in soft tissues around bones, but they look and act very much like Ewing sarcomas in bones. They are also known as *extraskkeletal* Ewing sarcomas.
- **Peripheral primitive neuroectodermal tumor (PPNET):** This rare childhood cancer also starts in bone or soft tissue and shares many features with Ewing sarcoma of bone and EOE. Peripheral PNETs that start in the chest wall are known as *Askin tumors*.

Researchers have found that the cells that make up Ewing sarcoma, EOE, and PPNET are very similar. They tend to have the same DNA (gene) abnormalities and share similar proteins, which are rarely found in other types of cancers. That's why these 3 cancers are thought to develop from the same type of cells in the body. There are slight differences among these tumors, but they're all [treated](#)<sup>4</sup> the same way.

Most Ewing tumors occur in the bones. The most common sites are:

- The pelvis (hip bones)
- The chest wall (such as the ribs or shoulder blades)
- The legs, mainly in the middle of the long bones

In contrast, osteosarcoma usually occurs at the ends of the long bones, especially around the knees. Extrasosseous Ewing tumors can occur almost anywhere.

Most Ewing tumors occur in children and teens, but they can also occur in adults. **This information focuses on the Ewing family of tumors in children and teens, but most of the information here (including much of the treatment information) applies to Ewing tumors in adults as well.**

## Other types of bone cancers

Several other types of cancers can start in the bones.

Osteosarcomas are the most common bone cancer in children and teens. They are described in [Osteosarcoma](#)<sup>5</sup>.

Most other types of bone cancers are usually found in adults and are rare in children. These include:

- Chondrosarcoma (cancer that develops from cartilage)
- Malignant fibrous histiocytoma
- Fibrosarcoma
- Chordoma
- Malignant giant cell tumor of bone

For more information on these cancers, see [Bone Cancer](#)<sup>6</sup>.

Many types of cancer that start in other organs of the body can spread to the bones. These are sometimes referred to as *metastatic bone cancers*, but they are not true bone cancers. For example, if a [rhabdomyosarcoma](#)<sup>7</sup> (a cancer that starts in muscle cells) spreads to the bones, it is still rhabdomyosarcoma and is treated like rhabdomyosarcoma. For more information, see [Advanced Cancer, Metastatic Cancer, and Bone Metastasis](#)<sup>8</sup>.

## Hyperlinks

1. [www.cancer.org/cancer/cancer-basics/what-is-cancer.html](http://www.cancer.org/cancer/cancer-basics/what-is-cancer.html)
2. [www.cancer.org/cancer/cancer-in-children.html](http://www.cancer.org/cancer/cancer-in-children.html)
3. [www.cancer.org/cancer/osteosarcoma.html](http://www.cancer.org/cancer/osteosarcoma.html)
4. [www.cancer.org/cancer/ewing-tumor/treating.html](http://www.cancer.org/cancer/ewing-tumor/treating.html)
5. [www.cancer.org/cancer/osteosarcoma.html](http://www.cancer.org/cancer/osteosarcoma.html)
6. [www.cancer.org/cancer/bone-cancer.html](http://www.cancer.org/cancer/bone-cancer.html)
7. [www.cancer.org/cancer/rhabdomyosarcoma.html](http://www.cancer.org/cancer/rhabdomyosarcoma.html)
8. [www.cancer.org/treatment/understanding-your-diagnosis/advanced-cancer.html](http://www.cancer.org/treatment/understanding-your-diagnosis/advanced-cancer.html)

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## Key Statistics for Ewing Tumors

About 1% of all childhood cancers are Ewing tumors. About 200 children and teens are diagnosed with Ewing tumors in the United States each year.

Most Ewing tumors occur in teens, but they can also affect younger children, as well as adults (mainly in their 20s and 30s).

Slightly more males than females develop these cancers. These tumors are much more common among whites, either non-Hispanic or Hispanic. This disease is very rare among African Americans, and it also seldom occurs in other racial groups.

Survival statistics for these tumors are discussed in [Survival Rates for Ewing Tumors, by Stage](#)<sup>1</sup>.

Visit the [American Cancer Society's Cancer Statistics Center](#)<sup>2</sup> for more key statistics.

### Hyperlinks

1. [www.cancer.org/cancer/ewing-tumor/detection-diagnosis-staging/survival-rates.html](http://www.cancer.org/cancer/ewing-tumor/detection-diagnosis-staging/survival-rates.html)
2. <https://cancerstatisticscenter.cancer.org/>

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American Cancer Society. *Cancer Facts & Figures 2020*. Atlanta, Ga: American Cancer Society; 2020.

Anderson ME, Randall RL, Springfield DS, Gebhart MC. Chapter 92: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

DeLaney TF, Hornicek FJ, Bahrami A. Epidemiology, pathology and molecular genetics of the Ewing sarcoma family of tumors. UpToDate. Accessed at [www.uptodate.com/contents/epidemiology-pathology-and-molecular-genetics-of-the-ewing-sarcoma-family-of-tumors](http://www.uptodate.com/contents/epidemiology-pathology-and-molecular-genetics-of-the-ewing-sarcoma-family-of-tumors) on February 27, 2018.

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# What's New in Ewing Tumor Research and Treatment?

Research on Ewing tumors is being done at many medical centers, university hospitals, and other institutions across the world.

## Understanding and diagnosing Ewing tumors

Scientists are developing new techniques to more accurately diagnose Ewing tumors. New [lab tests](#)<sup>1</sup> of tumor samples are being studied to see if they can help identify Ewing tumors and give more information on how well treatments might cure that particular tumor.

As researchers learn more about the changes inside Ewing tumor cells that make them different from normal cells, they hope to develop new treatments that take advantage of these changes (see below).

## Treatment

Researchers are looking to develop better treatments for Ewing tumors, as well as to find less toxic treatments for those that can be cured.

### Radiation therapy

Ewing tumors are very sensitive to [radiation therapy](#)<sup>2</sup>, but because of its possible side effects, it's most often used only if [surgery](#)<sup>3</sup> can't be done for some reason. Newer, more focused types of radiation therapy can help doctors treat tumors while lowering the dose of radiation to nearby healthy body tissues.

### Chemotherapy

The Children's Oncology Group, as well as many cancer centers and children's hospitals, are studying new chemotherapy combinations, which often include drugs such as topotecan, irinotecan, temozolomide, gemcitabine, docetaxel, and mithramycin (plicamycin).

Doctors are also trying to make the [currently used drugs](#)<sup>4</sup> more effective by changing the way they are given. For example, they have found that giving the standard VAC/IE (VDC/IE) chemo regimen more often – that is, every 2 weeks instead of every 3 weeks – seems to lower the chance of localized Ewing tumors coming back, without increasing the risk of serious side effects. This is often called *compressed chemotherapy*.

Researchers are also studying [high-dose chemotherapy with stem cell transplants](#)<sup>5</sup> in those with Ewing tumors that are unlikely to be cured with current treatments.

### Targeted therapy

As noted in [What Causes Ewing Tumors?](#)<sup>6</sup>, great progress is being made in understanding the changes in genes and chromosomes that cause Ewing tumors to form.

This knowledge has already been used to develop very sensitive lab tests to detect this cancer, and doctors are now studying how to best use these tests to guide the choice of treatment. It might also lead to new drugs that target these changes in Ewing tumor cells.

Some new drugs that target specific changes in Ewing tumor cells are already being tested. For example, TK216 is a drug that targets the main fusion protein in Ewing

tumor cells that is thought to help these cells grow. Early studies of this drug in people with Ewing tumors are now under way.

Also being studied in [clinical trials](#)<sup>7</sup> are drugs that target the insulin-like growth factor receptor-1 (IGF-1R), a protein on some cancer cells that causes them to grow. Early studies have found that drugs like this, such as ganitumab, can shrink some Ewing tumors and slow down the growth of others. So far, this benefit has been temporary in most cases. These drugs may work best when combined with other drugs.

Other newer drugs being studied for use against Ewing tumors include:

- Drugs that affect a tumor's ability to make new blood vessels, such as bevacizumab (Avastin)
- Drugs that target the mTOR protein, such as temsirolimus (Torisel) and everolimus (Afinitor)
- Drugs that target the PARP protein, such as olaparib (Lynparza) and niraparib (Zejula)
- Drugs called HDAC inhibitors, such as vorinostat (Zolinza)

## Immune therapy

A newer approach to treatment is to try to get the body's own immune system to recognize and attack the tumor cells. Some newer types of immune therapies, such as [checkpoint inhibitors](#)<sup>8</sup>, have shown a great deal of promise in treating other types of cancer, and some of these approaches are now being looked at for Ewing tumors. These treatments are still in the early stages of testing at this time.

## Hyperlinks

1. [www.cancer.org/cancer/ewing-tumor/detection-diagnosis-staging/how-diagnosed.html](http://www.cancer.org/cancer/ewing-tumor/detection-diagnosis-staging/how-diagnosed.html)
2. [www.cancer.org/cancer/ewing-tumor/treating/radiation-therapy.html](http://www.cancer.org/cancer/ewing-tumor/treating/radiation-therapy.html)
3. [www.cancer.org/cancer/ewing-tumor/treating/surgery.html](http://www.cancer.org/cancer/ewing-tumor/treating/surgery.html)
4. [www.cancer.org/cancer/ewing-tumor/treating/chemotherapy.html](http://www.cancer.org/cancer/ewing-tumor/treating/chemotherapy.html)
5. [www.cancer.org/cancer/ewing-tumor/treating/high-dose-chemo-stem-cell.html](http://www.cancer.org/cancer/ewing-tumor/treating/high-dose-chemo-stem-cell.html)
6. [www.cancer.org/cancer/ewing-tumor/causes-risks-prevention/what-causes.html](http://www.cancer.org/cancer/ewing-tumor/causes-risks-prevention/what-causes.html)
7. [www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html](http://www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html)
8. [www.cancer.org/treatment/treatments-and-side-effects/treatment-](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-)

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