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## About Osteosarcoma

### Overview and Types

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

- [What Is Osteosarcoma?](#)

### Research and Statistics

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

- [Key Statistics for Osteosarcoma](#)
  - [What's New in Osteosarcoma Research?](#)
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## What Is Osteosarcoma?

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 then spread to other areas of the body. To learn more about cancer and how it starts and spreads, see [What Is Cancer?](#)<sup>1</sup>

Osteosarcoma (also called **osteogenic sarcoma**) is the most common type of cancer that starts in the bones. The cancer cells in these tumors look like early forms of bone cells that normally help make new bone tissue, but the bone tissue in an osteosarcoma

is not as strong as that in normal bones.

Most osteosarcomas occur in children, teens, and young adults. Teens are the most commonly affected age group, but osteosarcoma can develop at any age. (For information about the differences between childhood cancers and adult cancers, see [Cancer in Children<sup>2</sup>](#).)

## Where does osteosarcoma start?

In children, teens, and young adults, osteosarcoma usually starts in areas where the bone is growing quickly, such as near the ends of the leg or arm bones:

- Most tumors develop in the **bones around the knee**, either in the lower part of the thigh bone (distal femur) or the the upper part of the shinbone (proximal tibia).
- The **upper arm bone close to the shoulder** (proximal humerus) is the next most common site.

Still, osteosarcoma can develop in any bone, including the bones of the pelvis (hips), shoulder, and jaw. This is especially true in older adults.

## Subtypes of osteosarcoma

Based on how the cancer cells look under the microscope, osteosarcomas can be classified as high grade, intermediate grade, or low grade. The grade of the tumor tells doctors how likely it is that the cancer will grow quickly and spread to other parts of the body.

### High-grade osteosarcomas

These are the fastest growing types of osteosarcoma. When seen with a microscope, they do not look like normal bone, and many of the cancer cells are in the process of dividing into new cells. Most osteosarcomas that occur in children and teens are high grade. There are many types of high-grade osteosarcomas (although the first 3 are the most common).

- Osteoblastic
- Chondroblastic
- Fibroblastic
- Small cell

- Telangiectatic
- High-grade surface (juxtacortical high grade)

Other high-grade osteosarcomas include:

- Pagetoid: a tumor that develops in someone with [Paget disease of the bone](#)<sup>3</sup>
- Extraskeletal: a tumor that starts in a part of the body other than a bone (but still makes bone tissue)
- Post-radiation: a tumor that starts in a bone that had once been exposed to radiation

### **Intermediate-grade osteosarcomas**

These uncommon tumors fall between high-grade and low-grade osteosarcomas. (They are usually [treated](#)<sup>4</sup> the same way as low-grade osteosarcomas.)

- Periosteal (juxtacortical intermediate grade)

### **Low-grade osteosarcomas**

These are the slowest-growing osteosarcomas. The tumors look more like normal bone and have few dividing cells when seen with a microscope.

- Parosteal (juxtacortical low grade)
- Intramedullary or intraosseous well differentiated (low-grade central)

The grade of the tumor plays a role in determining its stage and the type of treatment used. For more on staging, see [Osteosarcoma Stages](#)<sup>5</sup>.

### **Other types of bone tumors**

Several other types of tumors can start in the bones.

#### **Malignant (cancerous) bone tumors**

**Ewing tumors (Ewing sarcomas)** are the second most common bone cancer in

children. They are described in [Ewing Family of Tumors](#)<sup>6</sup>.

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

- Chondrosarcoma (cancer that develops from cartilage)
- Undifferentiated pleomorphic sarcoma (UPS) of bone, previously known as malignant fibrous histiocytoma (MFH) of bone
- Fibrosarcoma of bone
- Chordoma
- Malignant giant cell tumor of bone

For more information on these cancers, see [Bone Cancer in Adults](#)<sup>7</sup>.

Many types of cancer that start in other organs of the body, especially cancers in adults, can spread to the bones. These are sometimes referred to as **metastatic bone cancers**, but they are not true bone cancers. For example, prostate cancer that spreads to the bones is still prostate cancer and is treated like prostate cancer. For more information, see [Bone Metastasis](#)<sup>8</sup>.

### **Benign (non-cancerous) bone tumors**

Not all bone tumors are cancer. Benign bone tumors do not spread to other parts of the body. They are usually not life threatening, and surgery can often remove them completely. There are many types of benign bone tumors, including:

- Osteoma
- Chondroma
- Osteochondroma
- Eosinophilic granuloma of bone
- Non-ossifying fibroma
- Enchondroma
- Benign giant cell tumor of bone
- Lymphangioma

### **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/causes-risks-prevention/risk-factors.html](http://www.cancer.org/cancer/osteosarcoma/causes-risks-prevention/risk-factors.html)
4. [www.cancer.org/cancer/osteosarcoma/treating/by-extent.html](http://www.cancer.org/cancer/osteosarcoma/treating/by-extent.html)
5. [www.cancer.org/cancer/osteosarcoma/detection-diagnosis-staging/staging.html](http://www.cancer.org/cancer/osteosarcoma/detection-diagnosis-staging/staging.html)
6. [www.cancer.org/cancer/ewing-tumor.html](http://www.cancer.org/cancer/ewing-tumor.html)
7. [www.cancer.org/cancer/bone-cancer.html](http://www.cancer.org/cancer/bone-cancer.html)
8. [/content/cancer/en/treatment/understanding-your-diagnosis/bone-metastasis.html](http://content.cancer/en/treatment/understanding-your-diagnosis/bone-metastasis.html)

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## Key Statistics for Osteosarcoma

Osteosarcoma is not a common cancer. Each year, about 1,000 new cases of osteosarcoma are diagnosed in the United States. About half of these are in children and teens.

Most osteosarcomas occur in children, teens, and young adults between the ages of 10

and 30. Teens are the most commonly affected age group, but people of any age can develop osteosarcoma. About 1 in 10 osteosarcomas occur in people older than 60.

About 2% of childhood cancers are osteosarcomas, but they make up a much smaller percentage of adult cancers.

The prognosis (outlook) for people with osteosarcoma depends on many factors, including where the tumor is, if the cancer has already spread (metastasized) when it's first found, and the person's age. For more on this, see [Survival Rates for Osteosarcoma](#).<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/osteosarcoma/detection-diagnosis-staging/survival-rates.html](http://www.cancer.org/cancer/osteosarcoma/detection-diagnosis-staging/survival-rates.html)
2. [cancerstatisticscenter.cancer.org/](http://cancerstatisticscenter.cancer.org/)

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# What's New in Osteosarcoma Research?

Research on osteosarcoma is now being done at many medical centers, university hospitals, and other institutions around the world.

## Understanding osteosarcoma

Researchers are learning more about what makes osteosarcoma cells different from normal bone cells. Knowing more about the changes in osteosarcoma cells might eventually result in specific treatments based on these changes.

For example, researchers have found that osteosarcoma cells often have large amounts of a substance called GD2 on their surfaces. Drugs that target GD2 are already used to treat [neuroblastoma](#)<sup>1</sup> (another cancer often seen in children). Newer immunotherapy treatments that target GD2 are now being studied for use against osteosarcoma as well (see below).

Lab tests of the gene changes inside osteosarcoma cells might help predict the behavior of each tumor, such as how they will respond to certain types of chemotherapy or targeted therapy drugs. This type of testing is now being studied in clinical trials.

## Treatment

Many advances have been made in treating osteosarcoma in the past few decades. Still, more research is needed to learn how best to manage hard-to-treat osteosarcomas, such as those that have already spread when they are found. Many clinical trials are focusing on treating osteosarcoma using a variety of strategies.

## Surgery

The typical patterns of osteosarcoma growth and spread are much better understood now than they were. Newer imaging tests can also better define the extent of tumors.

These advances, along with sophisticated computer programs that help surgeons map out the best surgical approach before and during the operation (known as **computer-assisted tumor surgery**, or **CATS**), can help surgeons remove the cancer while sparing as much normal tissue as possible.

Some newer types of internal prostheses (man-made replacements for pieces of bone) can now be extended without the need for more surgery. This is especially important for children, who in the past often needed several operations to replace the prosthesis with a larger one as they grew.

## Radiation therapy

Osteosarcoma cells are not killed easily by radiation, so high doses are needed to have an effect. Because high doses can often cause unacceptable side effects, this has limited the use of [radiation therapy](#)<sup>2</sup>. Newer forms of radiation let doctors focus radiation more precisely on the tumor. Limiting the doses that reach nearby healthy tissues may allow higher doses to be used on the tumor itself.

**Intensity-modulated radiation therapy (IMRT)** is an example of an advanced form of therapy. In this technique, radiation beams are shaped to fit the tumor and aimed at it from several angles. The intensity (strength) of the beams can also be adjusted to limit the dose reaching nearby normal tissues. Many hospitals and cancer centers now use IMRT, especially for tumors in hard-to-treat areas such as the spine or pelvis (hip bones).

**Stereotactic radiosurgery (SRS)** gives a large (usually one-time) dose of radiation to a small tumor area. Once imaging tests have shown the exact location of the tumor, a thin beam of radiation is focused on the area from many different angles. The radiation is very precisely aimed so that it has as little effect on nearby tissues as possible. Sometimes doctors give the radiation in several smaller treatments to deliver the same or slightly higher dose. This is called **fractionated stereotactic radiotherapy**.

Another newer approach is to use radioactive particles instead of x-rays to deliver the radiation. One example is **conformal proton beam therapy**, which uses positive parts of atoms. Unlike x-rays, which release energy both before and after they hit their target, protons cause little damage to normal tissues they pass through and then release their energy after traveling a certain distance. Doctors can use this property to deliver more radiation to the tumor and to do less damage to nearby normal tissues. Proton beam therapy may be helpful for hard-to-treat tumors, such as those on the spine or pelvic bones, but only a limited number of centers in the United States offer this treatment at this time.

An even newer approach uses **carbon ions**, which are heavier than protons and cause more damage to cancer cells. This therapy is still in the earliest stages of development and is only available in a small number of centers around the world.

Doctors are also studying newer forms of **radioactive drugs** to treat osteosarcoma that has spread to many bones. One example is radium-223 (Xofigo), which works slightly differently than the other radioactive drugs now being used.

## Chemotherapy

Clinical trials are being done to determine the best combinations of [chemotherapy](#)<sup>3</sup> (chemo) drugs, as well as the best time to give them. Newer chemo drugs are being studied as well.

The [lungs](#)<sup>4</sup> are the most common place for osteosarcoma to spread. Inhaled forms of some chemo drugs (such as cisplatin) are being studied for patients whose cancer has spread to their lungs.

## Other forms of treatment

Chemo drugs are often helpful in treating osteosarcoma, but sometimes they don't work, or the cancer becomes resistant to them over time. Researchers are studying newer types of drugs that attack osteosarcoma cells in different ways.

## Immunotherapy drugs

Clinical trials are looking into ways to help the patient's own immune system recognize and attack the osteosarcoma cells. For example:

- Drugs called [immune checkpoint inhibitors](#)<sup>5</sup> can sometimes help the body's immune system recognize and attack cancer cells. These drugs have already been shown to be helpful against many types of cancer, and some of them are now being studied for use against osteosarcoma.
- An experimental immune-stimulating drug called **muramyl tripeptide** (also known as MTP or mifamurtide) has been shown to help some patients when added to chemotherapy.
- [Monoclonal antibodies](#)<sup>6</sup> are man-made versions of immune system proteins that attach to a specific target in the body, which can help the immune system find and destroy cancer cells. Antibodies directed against GD2 and other substances on osteosarcoma cells are now being tested in clinical trials.

- Researchers are also studying a newer form of immunotherapy known as [CAR-T cell therapy](#)<sup>7</sup> for osteosarcoma that is no longer helped by other treatments.

## Targeted therapy drugs

Doctors are also studying new medicines that target specific molecules on or in cancer cells. These are known as [targeted therapies](#)<sup>8</sup>.

Monoclonal antibodies, discussed above, can also be considered a type of targeted therapy. These antibodies attach to certain substances on cancer cells, which can kill them or help to stop their growth. An example is **dinutuximab** (Unituxin), an antibody that attaches to GD2, a substance that is important for cancer cell growth.

Many other targeted drugs are being studied for use against osteosarcoma, including drugs that affect a tumor's ability to make new blood vessels, such as sorafenib (Nexavar), pazopanib (Votrient), lenvatinib (Lenvima), and cabozantinib (Cabometyx).

## Drugs that affect the bones

Drugs that target bone cells called osteoclasts may also be useful against osteosarcoma:

- **Bisphosphonates** are a group of drugs that are already used to treat osteoporosis (bone thinning) and certain cancers that have spread to the bones. Some of these drugs, such as pamidronate and zoledronic acid, are now being studied for use in osteosarcoma as well.
- **Denosumab** is a monoclonal antibody that targets the RANKL protein, which normally helps bones grow. It is now being studied for use against osteosarcoma.

## Hyperlinks

1. [www.cancer.org/cancer/neuroblastoma.html](http://www.cancer.org/cancer/neuroblastoma.html)
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  8. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html)

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