



About Osteosarcoma

Overview and Types

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

- [What Is Osteosarcoma?](#)
- [What Are the Differences Between Cancers in Adults and Children?](#)

Research and Statistics

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

- [What Are the Key Statistics About Osteosarcoma?](#)
- [What's New in Osteosarcoma Research and Treatment?](#)

What Are the Differences Between Cancers in Adults and Children?

The types of cancers that develop in children and teens are often different from the types that develop in adults. Childhood cancers are often the result of DNA changes in cells that take place very early in life, sometimes even before birth. Unlike many cancers in adults, childhood cancers are not strongly linked to lifestyle or environmental risk factors.

There are exceptions, but childhood cancers tend to respond better to treatments such as chemotherapy. Children's bodies also tend to tolerate chemotherapy better than adults' bodies do. But cancer treatments such as chemotherapy and radiation therapy can have long-term side effects, so children who survive cancer will need careful

attention for the rest of their lives.

Since the 1960s, most children and teens with cancer have been treated at specialized centers designed for them. These centers offer the advantage of being treated by a team of specialists who know the differences between adult and childhood cancers, as well as the unique needs of children with cancer and their families. This team usually includes pediatric oncologists, surgeons, radiation oncologists, pathologists, pediatric oncology nurses, and nurse practitioners.

These centers also have psychologists, social workers, child life specialists, nutritionists, rehabilitation and physical therapists, and educators who can support and educate the entire family.

Most children with cancer in the United States are treated at a center that is a member of the Children's Oncology Group (COG). All of these centers are associated with a university or children's hospital. As we have learned more about treating childhood cancer, it has become even more important that treatment be given by experts in this area.

When a child or teen is diagnosed with cancer, it affects every family member and nearly every aspect of the family's life. You can read more about coping with these changes in our document [Children Diagnosed With Cancer: Dealing With Diagnosis](#).

- [References](#)

[See all references for Osteosarcoma](#)

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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 spread to other areas of the body. To learn more about how cancers start and spread, see [What Is Cancer?](#)

Osteosarcoma (also called *osteogenic sarcoma*) is a type of cancer that starts in the

bones. To understand osteosarcoma, it helps to know about bones and what they do.

About normal bones

Many people think of bones as just being part of the skeleton, like the steel girders that support a building. But bones actually do a number of different things.

- Some bones help support and protect our vital organs. Examples include the skull bones, breast bone (sternum), and ribs. These types of bones are often referred to as *flat bones*.
- Other bones, such as those in the arms and legs, make a framework for our muscles that helps us move. These are called *long bones*.
- Bones also make new blood cells. This is done in the soft, inner part of some bones called the *bone marrow*, which contains blood-forming cells. New red blood cells, white blood cells, and platelets are made in bone marrow.
- Bones also provide the body with a place to store minerals such as calcium.

Because bones are very hard and don't change shape at least once we reach adulthood we might not think of bones as being alive, but they are. Like all other tissues of the body, bones have many kinds of living cells. Two main types of cells in our bones help them stay strong and keep their shape.

- *Osteoblasts* help build up bones by forming the bone matrix (the connective tissue and minerals that give bone its strength).
- *Osteoclasts* break down bone matrix to prevent too much of it from building up, and they help bones keep their proper shape.

By depositing or removing minerals from the bones, osteoblasts and osteoclasts also help control the amount of these minerals in the blood.

Osteosarcoma

Osteosarcoma is the most common type of cancer that develops in bone. Like the osteoblasts in normal bone, the cells that form this cancer make bone matrix. But the bone matrix of an osteosarcoma is not as strong as that of normal bones.

Most osteosarcomas occur in children and young adults. Teens are the most commonly affected age group, but osteosarcoma can occur at any age.

In children and young adults, osteosarcoma usually develops in areas where the bone

is growing quickly, such as near the ends of the long bones. Most tumors develop in the bones around the knee, either in the distal femur (the lower part of the thigh bone) or the proximal tibia (the upper part of the shinbone). The proximal humerus (the part of the upper arm bone close to the shoulder) is the next most common site. However, 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

Several subtypes of osteosarcoma can be identified by how they look on x-rays and under the microscope. Some of these subtypes have a better prognosis (outlook) than others.

Based on how they 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 and spread to other parts of the body.

High-grade osteosarcomas: These are the fastest growing types of osteosarcoma. When seen under a microscope, they do not look like normal bone and have many cells 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
- Mixed
- 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](#)
- Extra-skeletal: a tumor that starts in a part of the body other than a bone
- Post-radiation: a tumor that starts in a bone that had once received radiation therapy

Intermediate-grade osteosarcomas: These uncommon tumors fall in between high-grade and low-grade osteosarcomas. (They are usually treated as if they are 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 under 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 the section [How Is Osteosarcoma Staged?](#)

Other types of bone tumors

Several other types of tumors can start in the bones.

Malignant (cancerous) bone tumors

Ewing tumors are the second most common bone cancer in children. They are described in our document [Ewing Family of Tumors](#).

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 our document [Bone Cancer](#).

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, prostate cancer that spreads to the bones is still prostate cancer and is treated like prostate cancer. For more information, see the document [Bone Metastasis](#).

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 can often be cured by surgery. There are

many types of benign bone tumors.

- Osteomas are benign tumors formed by bone cells.
- Chondromas are benign tumors formed by cartilage cells.
- Osteochondromas are benign tumors with both bone and cartilage cells.

Other benign bone tumors include eosinophilic granuloma of bone, non-ossifying fibroma, enchondroma, xanthoma, benign giant cell tumor of bone, and lymphangioma.

The rest of this document covers only osteosarcoma.

- [References](#)

[See all references for Osteosarcoma](#)

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What Are the Key Statistics About Osteosarcoma?

Osteosarcoma is not a common cancer. Each year, about 1,000 new cases of osteosarcoma are diagnosed in the United States. About 450 of these are in children and teens.

Most osteosarcomas occur in children and young adults between the ages of 10 and 30. Teens are the most commonly affected age group, but osteosarcoma can occur in people of any age. About 10% of all osteosarcomas occur in people over the age of 60.

Osteosarcomas account for about 2% of childhood cancers, but they make up a much smaller percentage of adult cancers.

The prognosis (outlook) for people with osteosarcoma depends on many factors, including the location of the tumor, whether the cancer has spread (metastasized) when it's first found, and the person's age. Statistics related to survival are discussed in the section [What Are the Survival Rates for Osteosarcoma?](#)

Visit the [American Cancer Society's Cancer Statistics Center](#) for more key statistics.

- [References](#)

[See all references for Osteosarcoma](#)

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What's New in Osteosarcoma Research and Treatment?

Research on osteosarcoma is now being done at medical centers, university hospitals, and other institutions across 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 that exploit 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](#) (another cancer often seen in children), and are now being studied for use against osteosarcoma.

Tests of gene changes called *gene expression profiling* might help predict the behavior of each tumor, such as how they will respond to certain types of chemotherapy. These are still being tested in clinical trials.

Treatment

Great 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

Doctors now have a much better understanding of the typical growth and spread of osteosarcomas than they did in the past. This, along with newer imaging tests that better define the extent of tumors, lets them plan surgeries to remove the cancer while sparing as much normal tissue as possible.

Some newer types of internal prostheses (man-made devices used to replace pieces of bone) can now be expanded 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. This has limited the use of radiation, because such high doses can often cause unacceptable side effects. Newer forms of radiation let doctors focus the radiation more precisely on the tumor. This limits the doses received by nearby healthy tissues and 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 the tumor from several angles. The intensity (strength) of the beams can also be adjusted to limit the dose reaching nearby normal tissues. This may let the doctor deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT, especially for tumors in hard-to-treat areas such as the spine or pelvis (hip bones).

A newer approach is to use radioactive particles instead of x-rays to deliver the radiation. One example uses protons, which are positive parts of atoms. Unlike x-rays, which release energy both before and after they hit their target, protons cause little damage to 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. As with IMRT, proton beam therapy may be helpful for hard-to-treat tumors, such as those on the spine or pelvic bones. The machines needed to make protons are expensive, and there are only a handful of them in the United States 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 few 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 might work better than the drugs now used.

Chemotherapy

Clinical trials are being done to determine the best combinations of chemotherapy (chemo) drugs, as well as the best time to give them. Newer chemo drugs are being studied as well.

The lungs 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. Early results have been promising.

Other new forms of treatment

Chemo drugs are often effective against 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. An experimental immunomodulating drug called *muramyl tripeptide* (also known as MTP or mifamurtide) has been shown to help some patients when added to chemotherapy.

Targeted therapy drugs: Doctors are also studying new medicines that target specific molecules on the cancer cells. These are known as *targeted therapies*. Some of these are man-made versions of immune system proteins, known as *monoclonal antibodies*. These antibodies attach to certain proteins on the cancer cell and help to stop the growth or kill the cancer cells. Examples now being studied include antibodies against the insulin-like growth factor receptor 1 (IGF-1R), a protein that may help cancer cells grow.

Other targeted drugs being studied for use against osteosarcoma include:

- Drugs that affect a tumor's ability to make new blood vessels, such as sorafenib (Nexavar) and pazopanib (Votrient).
- Drugs that target the mTOR protein, such as temsirolimus (Torisel) and everolimus (Afinitor).

Drugs that affect the bones: Other 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 bone. Some of these drugs, such as pamidronate and zoledronic acid, are now being studied for use in osteosarcoma as well. Another drug that affects bones, known as *saracatinib* (AZD0530), is also being studied.

- [References](#)

[See all references for Osteosarcoma](#)

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