



# About Bone Cancer

## Overview and Types

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

- [What is Bone Cancer?](#)

## Research and Statistics

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

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

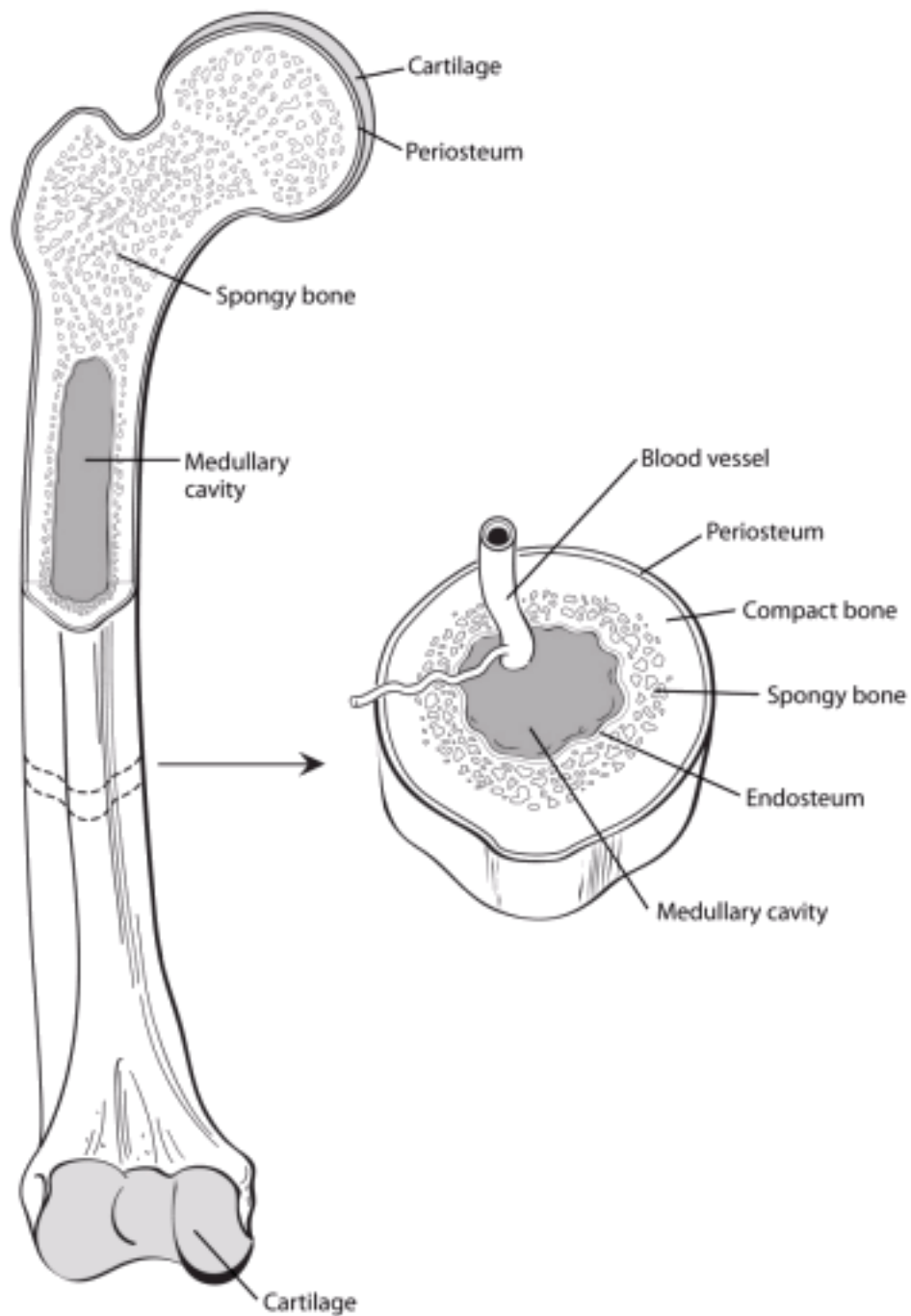
# What is Bone Cancer?

Bone cancer starts in the bone. 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?](#)

## Normal bone tissue

To understand bone cancer, it helps to understand a little about normal bone tissue.

*Bone* is the supporting framework of your body. Most bones are hollow. The outer part of bones is a network of fibrous tissue called *matrix* onto which calcium salts are laid down.



The hard outer layer of bones is made of compact (cortical) bone, which covers the lighter spongy (trabecular) bone inside. The outside of the bone is covered with a layer of fibrous tissue called *periosteum*. Some bones are hollow and have a space called the *medullary cavity* which contains the soft tissue called *bone marrow* (discussed below).

The tissue lining the medullary cavity is called *endosteum*. At each end of the bone is a zone of a softer form of bone-like tissue called *cartilage*.

Cartilage is softer than bone but more firm than most tissues. It is made of a fibrous tissue matrix mixed with a gel-like substance that does not contain much calcium.

Most bones start out as cartilage. The body then lays calcium down onto the cartilage to form bone. After the bone is formed, some cartilage may remain at the ends to act as a cushion between bones. This cartilage, along with ligaments and some other tissues connect bones to form a joint. In adults, cartilage is mainly found at the end of some bones as part of a joint. It is also seen at the place in the chest where the ribs meet the sternum (breastbone) and in parts of the face. The trachea (windpipe), larynx (voice box), and the outer part of the ear are other structures that contain cartilage.

Bone itself is very hard and strong. Some bone is able to support as much as 12,000 pounds per square inch. It takes as much as 1,200 to 1,800 pounds of pressure to break the femur (thigh bone). The bone itself contains 2 kinds of cells. The *osteoblast* is the cell that lays down new bone, and the *osteoclast* is the cell that dissolves old bone. Bone often looks as if it doesn't change much, but the truth is that it is very active. Throughout our bodies, new bone is always forming while old bone is dissolving.

In some bones the marrow is only fatty tissue. The marrow in other bones is a mixture of fat cells and blood-forming cells. The blood-forming cells produce red blood cells, white blood cells, and blood platelets. Other cells in the marrow include plasma cells, fibroblasts, and reticuloendothelial cells.

Cells from any of these tissues can develop into a cancer.

## Types of bone tumors

Most of the time when someone with cancer is told they have cancer in the bones, the doctor is talking about a cancer that has spread to the bones from somewhere else. This is called *metastatic cancer*. It can be seen in many different types of [advanced cancer](#), like breast cancer, prostate cancer, and lung cancer. When these cancers in the bone are looked at under a microscope, they look like the tissue they came from. For example, if someone has lung cancer that has spread to bone, the cells of the cancer in the bone still look and act like lung cancer cells. They do not look or act like bone cancer cells, even though they are in the bones. Since these cancer cells still act like lung cancer cells, they still need to be treated with drugs that are used for lung cancer.

For more information about metastatic bone cancer, please see [Bone Metastasis](#), as

well as information on the specific cancer ( [Breast Cancer](#), [Lung Cancer](#), [Prostate Cancer](#), etc.).

Other kinds of cancers that are sometimes called “bone cancers” start in the blood forming cells of the bone marrow not in the bone itself. The most common cancer that starts in the bone marrow and causes bone tumors is called [multiple myeloma](#). Another cancer that starts in the bone marrow is *leukemia*, but it is generally considered a blood cancer rather than a bone cancer. Sometimes lymphomas, which more often start in lymph nodes, can start in bone marrow. Multiple myeloma, lymphoma, and leukemia are not discussed in this document. For more information on these cancers, refer to the individual document for each.

A *primary* bone tumor starts in the bone itself. True (or primary) bone cancers are called *sarcomas*. Sarcomas are cancers that start in bone, muscle, fibrous tissue, blood vessels, fat tissue, as well as some other tissues. They can develop anywhere in the body.

There are several different types of bone tumors. Their names are based on the area of bone or surrounding tissue that is affected and the kind of cells forming the tumor. Some primary bone tumors are *benign* (not cancerous), and others are *malignant* (cancerous). Most bone cancers are sarcomas.

## **Benign bone tumors**

Benign tumors do not spread to other tissues and organs and so are not usually life threatening. They are generally cured by surgery. Types of benign bone tumors include:

- Osteoid osteoma
- Osteoblastoma
- Osteochondroma
- Enchondroma
- Chondromyxoid fibroma.

**These benign tumors are not discussed further here. This information is only about bone cancers.**

## **Malignant bone tumors**

**Osteosarcoma:** Osteosarcoma (also called *osteogenic sarcoma*) is the most common primary bone cancer. This cancer starts in the bone cells. It most often occurs in young people between the ages of 10 and 30, but about 10% of osteosarcoma cases develop

in people in their 60s and 70s. It is rare in middle-aged people, and is more common in males than females. These tumors develop most often in bones of the arms, legs, or pelvis. This cancer is not discussed in detail in this document, but is covered in [Osteosarcoma](#).

**Chondrosarcoma:**Chondrosarcoma is a cancer of cartilage cells. It is the second most common primary bone cancer. This cancer is rare in people younger than 20. After age 20, the risk of getting a chondrosarcoma goes up until about age 75. Women get this cancer as often as men.

Chondrosarcomas can develop anywhere there is cartilage. Most develop in bones such as the pelvis, leg bone or arm bone. Occasionally, chondrosarcoma will develop in the trachea, larynx, and chest wall. Other sites are the scapula (shoulder blade), ribs, or skull.

Benign (non-cancerous) tumors of cartilage are more common than malignant ones. These are called *enchondromas*. Another type of benign tumor that has cartilage is a bony projection capped by cartilage called an *osteochondroma*. These benign tumors rarely turn into cancer. There is a slightly higher chance of cancer developing in people who have many of these tumors, but this is still not common.

Chondrosarcomas are classified by grade, which measures how fast they grow. The grade is assigned by the pathologist (a doctor specially trained to examine and diagnose tissue samples under a microscope). The lower the grade, the slower the cancer grows. When a cancer is slow growing, the chance that it will spread is lower and so the outlook is better. Most chondrosarcomas are either low grade (grade I) or intermediate grade (grade II). High-grade (grade III) chondrosarcomas, which are the most likely to spread, are less common.

Some chondrosarcomas have distinctive features under a microscope. These variants of chondrosarcoma can have a different prognosis (outlook) than usual chondrosarcomas.

- *Dedifferentiated chondrosarcomas* start out as typical chondrosarcomas but then some parts of the tumor change into cells like those of a high-grade sarcoma (such as high grade forms of malignant fibrous histiocytoma, osteosarcoma, or fibrosarcoma). This variant of chondrosarcoma tends to occur in older patients and is more aggressive than usual chondrosarcomas.
- *Clear cell chondrosarcomas* are rare and grow slowly. They rarely spread to other parts of the body unless they have already come back several times in the original location.

- *Mesenchymal chondrosarcomas* can grow rapidly, but like Ewing tumor, are sensitive to treatment with radiation and chemotherapy.

**Ewing tumor:** Ewing tumor is the third most common primary bone cancer, and the second most common in children, adolescents, and young adults. This cancer (also called *Ewing sarcoma*) is named after the doctor who first described it in 1921, Dr. James Ewing. Most Ewing tumors develop in bones, but they can start in other tissues and organs. The most common sites for this cancer are the pelvis, the chest wall (such as the ribs or shoulder blades), and the long bones of the legs or arms. This cancer is most common in children and teenagers and is rare in adults older than 30. Ewing tumors occur most often in white people and are very rare among African Americans and Asian Americans. More detailed information about this cancer can be found in our document called [Ewing Family of Tumors](#).

**Malignant fibrous histiocytoma:** Malignant fibrous histiocytoma (MFH) more often starts in soft tissue (connective tissues such as ligaments, tendons, fat, and muscle) than in bones. This cancer is also known as *pleomorphic undifferentiated sarcoma*, especially when it starts in soft tissues. When MFH occurs in bones, it usually affects the legs (often around the knees) or arms. This cancer most often occurs in elderly and middle-aged adults and is rare among children. MFH mostly tends to grow locally, but it can spread to distant sites, like the lungs.

**Fibrosarcoma:** This is another type of cancer that develops more often in soft tissues than it does in bones. Fibrosarcoma usually occurs in elderly and middle-aged adults. Bones in the legs, arms, and jaw are most often affected.

**Giant cell tumor of bone:** This type of primary bone tumor has benign and malignant forms. The benign (non-cancerous) form is most common. Giant cell bone tumors typically affect the leg (usually near the knees) or arm bones of young and middle-aged adults. They don't often spread to distant sites, but tend to come back where they started after surgery (this is called *local recurrence*). This can happen several times. With each recurrence, the tumor becomes more likely to spread to other parts of the body. Rarely, a malignant giant cell bone tumor spreads to other parts of the body without first recurring locally.

**Chordoma:** This primary tumor of bone usually occurs in the base of the skull and bones of the spine. It develops most often in adults older than 30, and is about twice as common in men as in women. Chordomas tend to grow slowly and often do not spread to other parts of the body, but they often come back in the same area if they are not removed completely. The lymph nodes, the lungs, and the liver are the most common areas for secondary tumor spread.

## Other cancers that develop in bones

### Non-Hodgkin lymphomas

Non-Hodgkin lymphoma generally develops in lymph nodes but sometimes starts in the bone. Primary non-Hodgkin lymphoma of the bone is often a widespread disease because multiple sites in the body are usually involved. The outlook is similar to other non-Hodgkin lymphomas of the same subtype and stage. Primary lymphoma of the bone is given the same treatment as lymphomas that start in lymph nodes it is not treated like a primary bone sarcoma. For more information see [Non-Hodgkin Lymphoma](#).

### Multiple myelomas

Multiple myeloma almost always develops in bones, but doctors do not consider it a primary bone cancer because it develops from the plasma cells of the bone marrow (the soft inner part of some bones). Although it causes bone destruction, it is no more a bone cancer than is leukemia. It is treated as a widespread disease. At times, myeloma can be first found as a single tumor (called a *plasmacytoma*) in a single bone, but most of the time it will spread to the marrow of other bones. For more information see [Multiple Myeloma](#).

- [References](#)

[See all references for Bone Cancer](#)

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## What Are the Key Statistics About Bone Cancer?

The American Cancer Society's estimates for cancer of the bones and joints for 2017 are:

- About 3,260 new cases will be diagnosed.
- About 1,550 deaths from these cancers are expected.

Primary cancers of bones account for less than 0.2% of all cancers.

In adults, over 40% of primary bone cancers are chondrosarcomas. This is followed by osteosarcomas (28%), chordomas (10%), Ewing tumors (8%), and malignant fibrous histiocytoma/fibrosarcomas (4%). The remainder of cases are several rare types of bone cancers.

In children and teenagers (those younger than 20 years), [osteosarcoma](#) (56%) and [Ewing tumors](#) (34%) are much more common than chondrosarcoma (6%).

Chondrosarcomas develop most often in adults, with an average age at diagnosis of 51. Less than 5% of cases occur in patients younger than 20.

Chordomas are also more common in adults. Less than 5% of cases occur in patients younger than 20.

Both osteosarcomas and Ewing tumors occur most often in children and teens.

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

- [References](#)

[See all references for Bone Cancer](#)

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## What's New in Bone Cancer Research and Treatment?

Research on bone cancer is now being done at many medical centers, university hospitals, and other institutions across the nation. There are several ongoing clinical trials focusing on bone cancer.



# Chemotherapy

Some clinical trials are looking into ways to combine [surgery](#), [radiation therapy](#), and [chemotherapy](#) (chemo), and drugs known as [targeted therapy](#). One study found that the combination of the chemo drug cyclophosphamide (Cytoxan) and the targeted drug sirolimus can help stop chondrosarcomas from growing for a time.

Some are testing new chemo drugs.

# Targeted therapy

[Targeted therapy](#) drugs work differently from standard chemo. These drugs target certain genes and proteins in cancer cells.

One example of targeted therapy is the drug imatinib (Gleevec), which targets certain proteins made by the cancer cells in chordomas. Adding another drug to imatinib, such as the targeted therapy drug sirolimus (Rapamune<sup>®</sup>) or the chemo drug cisplatin helps stop the growth of chordomas when imatinib stops working. Another drug, panobinostat, is being studied in combination with imatinib to treat chordoma.

Lapatinib (Tykerb<sup>®</sup>) is another targeted drug that may be useful in treating chordoma. In one study of patients with tumors that had too many copies of the *EGFR* gene and/or too much EGFR protein, it helped shrink tumors and stop them from growing for a time.

Some chordomas show strong expression of parts of an insulin-like growth factor pathway. This has led to studying antibodies against the insulin-like growth factor receptor 1 (IGF-1R) in chordoma patients.

Studies of other targeted drugs are going on right now, such as nilotinib (Tasigna) and dasatinib (Sprycel) in chordoma, and pazopanib (Votrient<sup>®</sup>), everolimus (Afinitor<sup>®</sup>), and vismodegib (GDC-0449) in chondrosarcoma.

# Radiation

The most common type of radiation used to treat cancer uses beams of x-rays. Proton beam radiation uses particles made up of protons (protons are small positively charged particles that are part of atoms). Another much less common form of particle radiation that can be used to treat chordomas and chondrosarcomas is carbon ion radiation. This can be helpful in treating tumors of the skull base, but is only available in a few centers

worldwide.

## Genetics

In addition to [clinical trials](#), researchers are making progress in learning about the causes of bone tumors. For example, changes to a certain part of chromosome 6 have been found in chordomas. Changes to the *COL2A1* gene, which codes for a major form of collagen found in cartilage, have been found in many chondrosarcomas. Hopefully more information about the DNA changes that cause bone cancers will eventually lead to treatments aimed at these gene defects.

- [References](#)

[See all references for Bone Cancer](#)

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