



# 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.

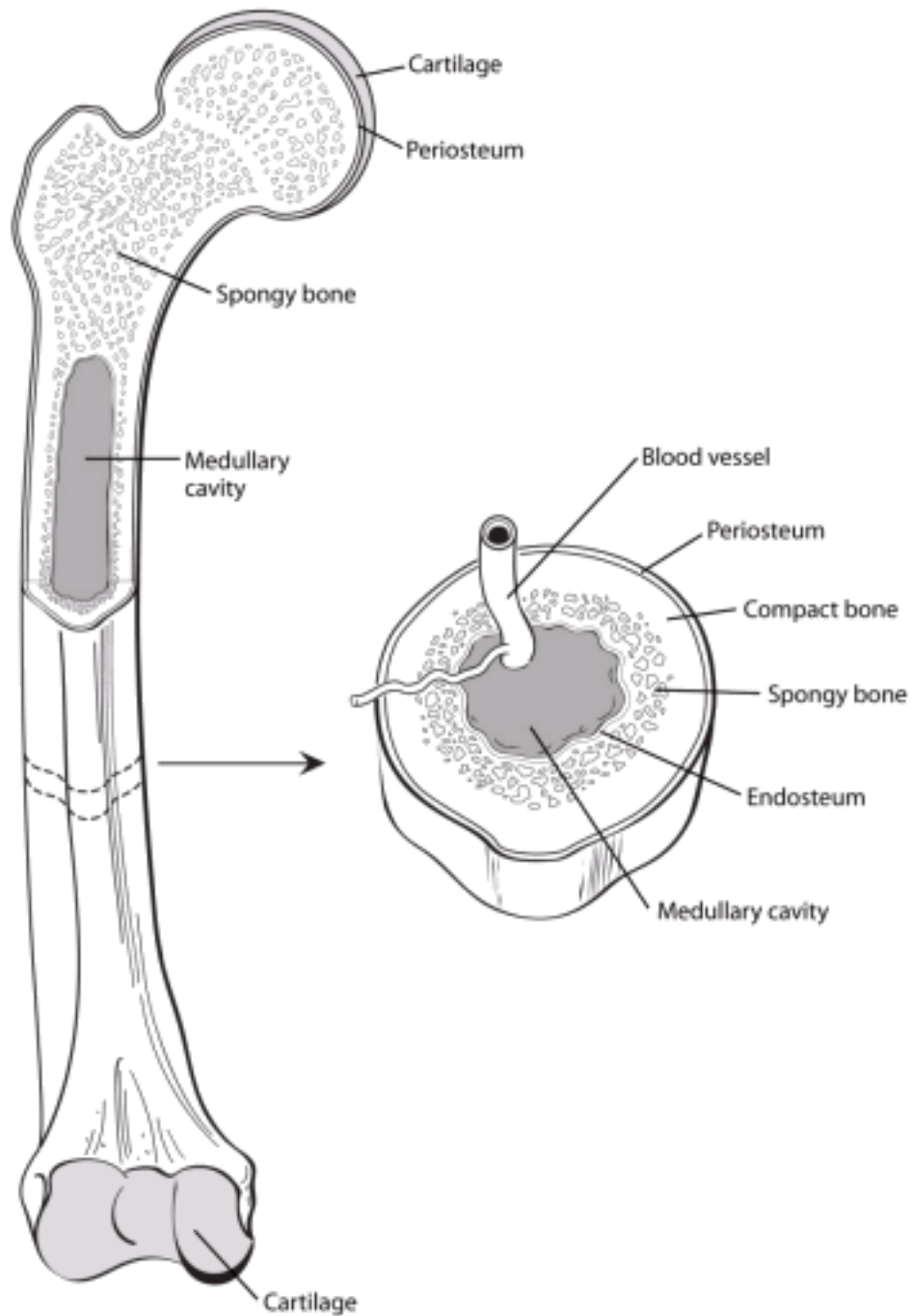
- [Key Statistics About Bone Cancer](#)
- [What's New in Bone Cancer Research?](#)

# What Is Bone Cancer?

Bone cancer is very rare in adults. It starts in the cells that make up the bone. Cancer starts when cells begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other parts 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 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 fibrous tissue called **periosteum**. Hollow bones have a space called the **medullary cavity** which contains

the soft, spongy 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's made of a fibrous tissue matrix mixed with a gel-like substance that doesn't 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, cartilage may remain at the ends to act as a cushion between bones. This cartilage, along with ligaments and other tissues connect bones to form a joint. In adults, cartilage is mainly found at the end of some bones that are part of a joint.

Cartilage is also 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 is very hard and strong. Some bone is able to support as much as 12,000 pounds of pressure per square inch. It takes as much as 1,200 to 1,800 pounds of pressure to break the femur (thigh bone).

Bone itself contains 2 kinds of cells.

- The **osteoblast** is the cell that lays down new bone
- The **osteoclast** is the cell that dissolves old bone.

Bone often looks as if it doesn't change much, but it's actually very active. New bone is always forming while old bone is dissolving.

In some bones the marrow is only fatty tissue. In other bones it's a mixture of fat cells and blood-forming cells. The blood-forming cells make red blood cells, white blood cells, and blood platelets. There are other cells in the bone marrow, too, such as plasma cells and fibroblasts.

Any of these bone cells can develop into cancer.

## **Types of bone tumors**

### **Bone tumors that are not cancer**

Some tumors that start in the bone are benign (not cancer). Benign tumors do not spread to other tissues and organs and are not usually life threatening. They often can

be cured with surgery. Types of benign bone tumors include:

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

Benign tumors are not discussed further here.

## **Bone metastases**

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 happen with many different types of [advanced cancer](#), like [breast](#) cancer, [prostate](#) cancer, and [lung](#) cancer. When the cancer cells in the bone are looked at under a microscope, they look just like the tissue they came from.

So, if someone has lung cancer that has spread to bone, the cancer cells in the bone look and act like lung cancer cells. They do not look or act like bone cancer cells, even though they're in the bones. Because these cancer cells still act like lung cancer cells, they need to be treated with drugs that are used for lung cancer.

To learn more about this, see [Bone Metastasis](#).

## **Blood cancers**

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](#). Sometimes [lymphomas](#), which more often start in [lymph nodes](#), can start in bone marrow. These blood cancers are not discussed here.

## **Bone cancers**

**True (or primary) bone tumors start in the bone itself and are called sarcomas.** These are malignant tumors, which means they're cancer.

Sarcomas start in bone, muscle, fibrous tissue, blood vessels, fat tissue, as well as some other tissues. They can develop anywhere in the body. They're covered below.

## **Malignant bone tumors**

There are many different kinds of primary bone cancer. They're named based on the part of the bone or nearby tissue that's affected and the kind of cells forming the tumor. Some are quite rare.

### **Osteosarcoma**

**Osteosarcoma (also called osteogenic sarcoma) is the most common primary bone cancer.** It 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's 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 type of bone cancer is covered in [Osteosarcoma](#).

### **Chondrosarcoma**

Chondrosarcoma starts in cartilage cells. It's the second most common primary bone cancer. It's 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 start anywhere there's cartilage. Most develop in bones like the pelvis, legs, or arms . Sometimes chondrosarcoma starts in the trachea, larynx, or chest wall. Other sites are the scapula (shoulder blade), ribs, or skull.

Benign (not cancer) tumors are more common in the cartilage than malignant ones. These are called enchondromas. Another type of benign cartilage tumor is a bony projection capped by cartilage called an osteochondroma. These benign tumors rarely turn into cancer. People who have many of these tumors have a slightly higher chance of developing cancer, but this isn't 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 with 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, 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 which can be seen with a microscope. These sub-types of chondrosarcoma often have a different prognosis (outlook):

- **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 type of chondrosarcoma tends to develop in older patients and grows faster than usual chondrosarcomas.
- **Clear cell chondrosarcomas** are rare and grow slowly. They seldom 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 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, teens, and young adults. It's rare in adults older than 30. This cancer (also called **Ewing sarcoma**) is named after Dr. James Ewing, who first described it in 1921. 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. Ewing tumors occur most often in white people and are very rare among African Americans and Asian Americans. More information can be found in [Ewing Family of Tumors](#).

## Malignant fibrous histiocytoma

Malignant fibrous histiocytoma (MFH) most often starts in soft tissue (connective tissues such as ligaments, tendons, fat, and muscle); it's rare 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. It's quite rare in 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 (not cancer) and malignant forms. The benign form is most common. Giant cell bone tumors typically affect the legs (usually near the knees) or arms of young and middle-aged adults. They don't often spread to distant sites, but after surgery tend to come back where they started . (This is called local recurrence.) This can happen many 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. It's 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. 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 tumor spread.

## **Other cancers that develop in bones**

Other cancers can be found in the bones, but they don't start in the actual bone cells. They are not treated like primary bone cancer.

## **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 many bones 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's not treated like a primary bone sarcoma. For more information see [Non-Hodgkin Lymphoma](#).

## **Multiple myelomas**

Multiple myeloma almost always develops in bones, but it's not a primary bone cancer

because it starts in the plasma cells of the bone marrow (the soft inner part of some bones). Although it causes bone destruction, it's no more a bone cancer than leukemia is. It's 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. See [Multiple Myeloma](#).

- [References](#)

[See all references for Bone Cancer](#)

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## Key Statistics About Bone Cancer

The American Cancer Society's estimates for cancer of the bones and joints for 2018 are:

- About 3,450 new cases will be diagnosed
- About 1,590 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](#)

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[See all references for Bone Cancer](#)

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## What's New in Bone Cancer Research?

Research on [bone cancer](#) is now being done at many medical centers, university hospitals, and other institutions around the world. There are many [clinical trials](#) focusing on bone cancer.

Because primary bone cancer is rare in adults, it's been hard to study well. Most experts agree that treatment in a clinical trial should be considered, especially for people with [advanced](#) bone cancers (those that come back after treatment, don't respond to treatment, and/or spread to other parts of the body). This way people can get the best treatment available now and may also get the treatments that are thought to be even better.

## Treatment

Some clinical trials are looking into ways to combine [surgery](#), [radiation therapy](#), and

[chemotherapy](#) (chemo), and drugs known as [targeted therapy](#) to treat these cancers.

## Chemotherapy

Some studies are testing new chemo drugs. Researchers are also looking for new, and maybe better, ways to use the drugs we have. For instance, doctors are studying whether adding a bisphosphonate called zoledronic acid (Zometa) to the bone cement used to fill in the space left after removing a giant cell tumor might decrease the chance that the tumor will come back in that place.

Another area of interest is long-term chemotherapy side effects. Bone cancers are some of the more common cancers in young people, and doctors are trying to learn more about how the chemo drugs used might affect the way the brain develops and works as survivors grow older.

## Targeted therapy

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

A huge area of primary bone cancer research is learning more about the genetic changes in these cancer cells. Researchers are using existing drugs and developing new targeted drugs that focus on these gene changes. It's hoped that these drugs can change the cancer's ability to grow and spread, providing a new and better way to treat these tumors.

For instance, researchers have found that some giant cell tumors that have low levels of certain genes (called microRNA genes) are able to grow and spread faster. MicroRNA changes have also been found in chondrosarcomas. Tests that find these microRNA changes may be helpful in diagnosing these tumors. Finding drugs that target these genes might also prove to be a possible treatment.

One targeted therapy drug, nivolumab (Opdivo), is already used to treat other kinds of cancer. Doctors are trying to find out the best dose to use. They are looking at whether combining it with other treatments might slow tumor growth and help people with advanced sarcomas live longer.

The targeted therapy drug dasatinib (Sprycel) is also used to treat other cancers. Early studies have suggested it may help treat chondrosarcomas, both alone and combined with chemo. Chondrosarcomas seldom respond to chemo or radiation, so targeted therapy drugs may lead to new treatments for these hard-to-treat cancers.

Denosumab (Prolia or Xgeva) is another targeted therapy being tested to see if it can help control giant cell tumor of the bone and/or keep it from coming back after treatment. It's also been shown to help keep giant cell tumors from coming back after surgery.

Overall, results of targeted therapy research are not yet clear. More research is needed in this area, and many clinical trials are testing these treatments.

## Radiation

The most common type of radiation used to treat cancer uses beams of x-rays. Doctors are looking for better types of radiation. Proton beam radiation uses particles made up of protons. (Protons are small positively charged particles that are part of atoms.) Proton radiation is already used to treat bone tumors near very sensitive organs, like the brain or the spine. It's being tested on tumors in other parts of the body, too. And as advances make this treatment even more precise and more widely available, it may be found to work better in treating bone tumors.

Another much less common form of particle radiation that has been used to treat chordomas and chondrosarcomas is carbon ion radiation. This may be helpful in treating tumors that do not respond to available treatments, but a lot more research is needed. This treatment is only available in 10 centers worldwide, and there are no carbon ion radiation facilities in North America as of 2017.

## Genetics

In addition to [clinical trials](#), researchers are making progress in learning about the causes of bone tumors. For example, changes to the T gene have been found in a few families where more than one member has a chordoma. This might help doctors find specific gene changes that might put a person at higher risk for this type of bone cancer.

Other gene changes found in giant cell tumors may help doctors find better ways to both diagnose and treat these tumors.

Scientists hope that learning more about the DNA changes that cause bone cancers will also lead to better treatments that might be aimed at these gene defects.

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