National Comprehensive Cancer Network®

2026

NCCN Guidelines for Patients®

Cancer care recommendations from leading experts at the National Comprehensive Cancer Network® (NCCN®)

Bone Cancer



Presented with support from NCCN FOUNDATION® Guiding Treatment. Changing Lives.

Available online at NCCN.org/patientguidelines



NCCN Guidelines for Patients®

The essential guide for people facing cancer.

Based on care recommendations from leading cancer experts.

Explains high-quality cancer care provided at state-of-the-art cancer centers.

Reviewed and revised every year.

Did you know that top cancer centers across the United States work together to improve cancer care? This alliance of leading cancer centers is called the National Comprehensive Cancer Network® (NCCN®).

Because cancer care is always evolving, NCCN develops and frequently updates evidence-based cancer care recommendations used by health care providers worldwide. These recommendations are known as the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®).

The NCCN Guidelines for Patients plainly explain these expert recommendations, so you can talk with your care team about the best care for you.

These NCCN Guidelines for Patients are based on the NCCN Guidelines® for Bone Cancer, Version 1.2026 — September 11, 2025

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Contents

4 Chapter 1
About bone cancer

8 Chapter 2
Testing for bone sarcoma

15 Chapter 3
Types of treatment

26 Chapter 4
Osteosarcoma

32 Chapter 5
Ewing sarcoma

38 Chapter 6
Chondrosarcoma

43 Chapter 7
Chordoma

47 Chapter 8
Giant cell tumor of bone

52 Chapter 9
Other resources

56 Words to know

58 NCCN Contributors

59 NCCN Cancer Centers

60 Index

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1
About bone cancer

5 What is bone cancer?

6 What is bone?

6 How is bone cancer treated?

7 How can I get the best care?

Bone cancer includes a group of cancers that start in the bone or cartilage. This is called primary bone cancer and differs from other types of cancer that can spread to the bone. This chapter presents an overview of bone cancer.

What is bone cancer?

Bone cancer is cancer that starts in the bone or cartilage. This is called primary bone cancer and differs from other types of cancer that can spread to the bone.

Cancer cells don't behave like normal cells. They develop genetic changes that allow them to multiply and make many more cancer cells. The cancer cells crowd out and overwhelm normal cells.

Cancer cells also survive much longer than normal cells do. They can replace many normal cells and cause organs to stop working properly. This can end up harming the body.

Bone cancers that start in another part of the body (such as the breast, lungs, or kidney) and travel to the bones may be called **secondary bone malignancies** or **metastatic cancer to bone**.

Metastatic cancers to bone behave very differently from primary bone cancers.

Sarcomas

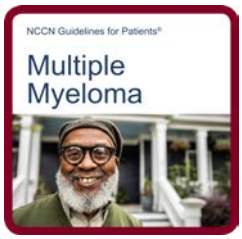
Sarcomas are cancers that start in the connective tissues of the body. The connective tissues include bone, cartilage, and the soft tissues of the limbs and body wall. There are 2 main types of sarcomas:

- Soft tissue sarcomas
- Bone sarcomas

Soft tissue includes fat, muscles, nerves, tendons, blood and lymph vessels, and other supportive tissues of the body. Soft tissue sarcomas are more common than bone sarcomas. Bone sarcomas, such as osteosarcomas, start in the cells that form bone.

For more information, see *NCCN Guidelines for Patients: Soft Tissue Sarcoma* and *NCCN Guidelines for Patients: Multiple Myeloma, available* at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.





Types of bone sarcomas

A bone tumor may be cancerous (malignant) or non-cancerous (benign). A bone sarcoma is an abnormal, malignant growth of cells within a bone. Bone sarcoma often occurs in areas of the bone that grow rapidly.

This guide will discuss:

- **Osteosarcoma** – usually starts in cells in the areas of the bone that grow rapidly. Read more in *Chapter 4: Osteosarcoma*.
- **Ewing sarcoma** – starts in the bone marrow and commonly occurs in the shaft of long bones or flat bones like the hip bone or shoulder blade. Read more in *Chapter 5: Ewing sarcoma.*
- **Chondrosarcoma** – starts in the cells that form cartilage. Cartilage is a tough, flexible tissue that covers the ends of bones to form a smooth surface that makes a joint. Also, cartilage gives structure to the nose, ears, larynx, and other parts of the body. Read more in *Chapter 6: Chondrosarcoma.*
- **Chordoma** – starts in cells that form the spine and vertebrae. Chordomas are usually found in the lower back or at the base of the skull. Read more in *Chapter 7: Chordoma.*
- **Giant cell tumor of bone (GCTB)** – is an overgrowth of cells that extends into the area of the bone that widens to form a joint. Read more in *Chapter 8: Giant cell tumor of bone.*

Osteosarcoma, Ewing sarcoma, and chondrosarcoma are the most common types of bone cancer.

What is bone?

Bone is a living tissue made of specialized cells and collagen. Bone tissue also contains blood vessels, nerves, proteins, vitamins, and minerals.

A fully developed human body has 206 bones. Bones provide structure and support, protect organs, create blood cells, store chemicals the body needs, and produce hormones. Bone is light yet strong and continuously remodels throughout your life.

How is bone cancer treated?

For chondrosarcoma, treatment is usually surgery to remove the tumor. Chordoma treatment options include surgery or radiation therapy. Ewing sarcoma treatment begins with drug therapy and includes surgery and/or radiation.

Giant cell tumor of bone (GCTB) is usually not cancer, but needs aggressive treatment to prevent bone damage.

Osteosarcoma is treated with a combination of medical and surgical therapies.

How can I get the best care?

Advocate for yourself. You have an important role to play in your care. Many people feel more satisfied when they actively take part in planning their cancer care.

The NCCN Guidelines for Patients will help you play a larger role in your care. Discuss the recommendations in this guide with your care team. Ask questions about your options and share your goals and concerns.

Don't know what to ask? You're not alone. That's why we include suggested questions to ask at the end of chapters.

Keep reading to find the best care for you.

How this guide can help you

Making decisions about cancer care is stressful. There's a lot to learn, and you don't know what the future holds.

Use this guide to get the information and support you need.

Patients, doctors, and other health care professionals trust the NCCN Guidelines for Patients. This guide uses clear, everyday language to explain current cancer care recommendations made by respected experts in the field. Their recommendations are based on the latest research and practices at leading cancer centers.

Your health is unique to you, so your cancer care should be, too. As you read this guide, you'll learn which treatments are likely to provide the best results for you. And you'll be better prepared to talk with your care team.

2
Testing for bone sarcoma

9 General health tests

10 Blood tests

11 Imaging tests

12 Biopsy

13 Genetic cancer risk testing

13 Biomarker testing

14 Key points

14 Questions to ask

Treatment planning starts with testing. Accurate testing is needed to diagnose and treat primary bone cancer. This chapter presents an overview of possible tests you might receive and what to expect.

Bone sarcoma is very rare. Many health care providers are not experts in diagnosing or treating it. Because of this, you should try to find a healthcare professional, treatment center, or hospital that has experience in your type of bone sarcoma.

Use the resources on page 52 of this book to help you find an experienced health care professional and treatment center.

You'll have general health tests first, followed by more specific tests. Some of these general health tests are described next.

General health tests

While the results of these tests might be available to you through your patient portal or patient access system, you should wait to discuss these results with your care team.

Medical history

A medical history is a record of all health issues and treatments you've had in your life. Be prepared to list any illness or injury and when it happened. Tell your care team about anything you're experiencing, as it may be a symptom of a deeper issue.

Bring a list of old and new medicines and any over-the-counter (OTC) medicines, herbal remedies, or supplements you take. Some supplements interact with and affect medicines that your care team may prescribe.

Family history

Your care team will ask about the health history of family members who are blood relatives. This is called a family history.

Ask family members on both sides of your family about their health issues like heart disease, cancer, and diabetes, and at what age they were diagnosed.

Guide 1
Tests for bone sarcoma

Medical history and physical exam
X-ray of primary site
MRI scan
CT scan
PET scan
Bone marrow biopsy
Biomarker testing
Complete blood count
Comprehensive metabolic panel
Alkaline phosphatase (ALP)
Lactate dehydrogenase (LDH)

For family members with cancer, try to find out the specific type of cancer or where the cancer started, if it is in multiple locations, and if the family member had genetic testing.

Physical exam

A member of your care team will perform a thorough physical exam of your body. This exam includes checking vital signs, such as heart rate and blood pressure. Your provider may also gently press on areas of your body, feeling for swollen organs or lymph nodes.

Performance status

Performance status is a rating of a person's general level of fitness and ability to perform daily self-care tasks. Your performance status can affect the choice of treatment.

Blood tests

While there are no blood tests currently available that can diagnose bone sarcomas, blood tests check for signs of other diseases and how well organs are working. They require a sample of your blood, which is removed through a needle placed into a vein in your arm. If your veins are difficult to find, let the lab technician know and they may have a device that can help find your veins. This way you'll get fewer pokes from the needle and it's less stressful.

One blood draw can check all of the following components in your blood.

Alkaline phosphatase

Alkaline phosphatase (ALP) is an enzyme found in the blood. High levels of ALP can be a sign of bone tumors, liver disorders, or pregnancy. A bone scan might be performed if you have high levels of ALP.

Complete blood count

A complete blood count measures the levels of red blood cells, white blood cells, and platelets in your blood. Red blood cells carry oxygen throughout your body, white blood cells fight infection, and platelets control bleeding.

Comprehensive metabolic panel

A comprehensive metabolic panel measures substances in your blood. It provides important information about how well your kidneys and liver are working, among other things.

Calcium is needed for healthy teeth, bones, and other body tissues. A comprehensive metabolic panel will look for higher than normal calcium levels in your blood, called hypercalcemia. Bone damage from cancer can cause your bones to release excess calcium into the bloodstream, which can lead to illness and other health issues.

Lactate dehydrogenase

Lactate dehydrogenase (LDH) is an enzyme found in most cells. An elevated level of LDH is a sign that cell damage has occurred. While fast-growing cells such as tumor cells release LDH, many other illnesses also raise LDH levels.

Imaging tests

Imaging tests take pictures of the inside of your body to look for cancer. A medical professional called a radiologist will look at the test and send a report to your care team.

X-ray

X-rays are a type of radiation. In small doses, they're used to make pictures of the inside of the body.

Bone scan

In a bone scan, a special camera will take pictures of a substance in your bones as it moves through your body. A bone scan uses a radiotracer, a substance that releases small amounts of radiation. Before the pictures are taken, the tracer will be injected into your vein. It can take a few hours for the tracer to enter your bones.

Areas of bone damage take up more radiotracer than healthy bone and will show up as bright spots on the pictures. Bone damage can be caused by cancer; cancer treatment; previous injuries; or other health problems, such as arthritis.

The scan takes about 30 minutes. You'll be asked to hold still during the scan. At one point, the scanner will be less than 1 inch from your face. Talk to your care team about what to expect.

CT and MRI

CT, also called a CAT scan, takes many x-rays of the same body part from different angles. The images are combined using a computer to make a series of cross-sectional pictures of the inside of the body.

MRI uses radio waves and powerful magnets to take pictures of areas inside of the body. Unlike CT, it doesn't use radiation. If you have any metal in your body, tell your radiation tech before having an MRI. Most metal is safe for MRI, but you should inform the MRI technicians so they can determine if there are any safety concerns before having the test completed.

The choice of CT versus MRI depends on the type and location of the tumor. For example, MRI is preferred for tumors in the brain, spine, liver, and extremities. Both CT and MRI can provide details about the size of the tumor and how close it is to nearby organs, nerves, and blood vessels.

A radiologist is an expert at interpreting medical scans. They will review the images and write a report. The radiologist will send this report to your health care provider, who will discuss the results with you.

Contrast

Before or during a CT or MRI, you might be given a contrast agent through a needle in your arm. Contrast is a substance that changes how tissues look in pictures.

PET

Positron emission tomography (PET) scans use a radioactive substance called a tracer or isotope. This substance accumulates in places that use a lot of energy in your body.

Most cancer cells, including sarcoma cells, use much more energy than normal cells and show up as bright spots on PET scans. However, not all bright spots are cancer. Not all cancers appear on PET scans.

PET is usually combined with CT scans to provide additional helpful information.

Biopsy

A biopsy is the removal of a sample of tissue from your body for testing. A diagnosis of bone cancer is confirmed using a biopsy. The biopsy should be performed by an experienced surgeon at the center where you'll receive treatment.

The preferred type of biopsy for bone cancer is a core needle biopsy. It removes tissue with a wide, hollow needle.

Another type, called an incisional biopsy or open biopsy, removes a small amount of tissue through a cut in the skin or body.

Bone marrow tests

Bone marrow tests might be done in certain cases.

There are 2 bone marrow tests that are often done at the same time:

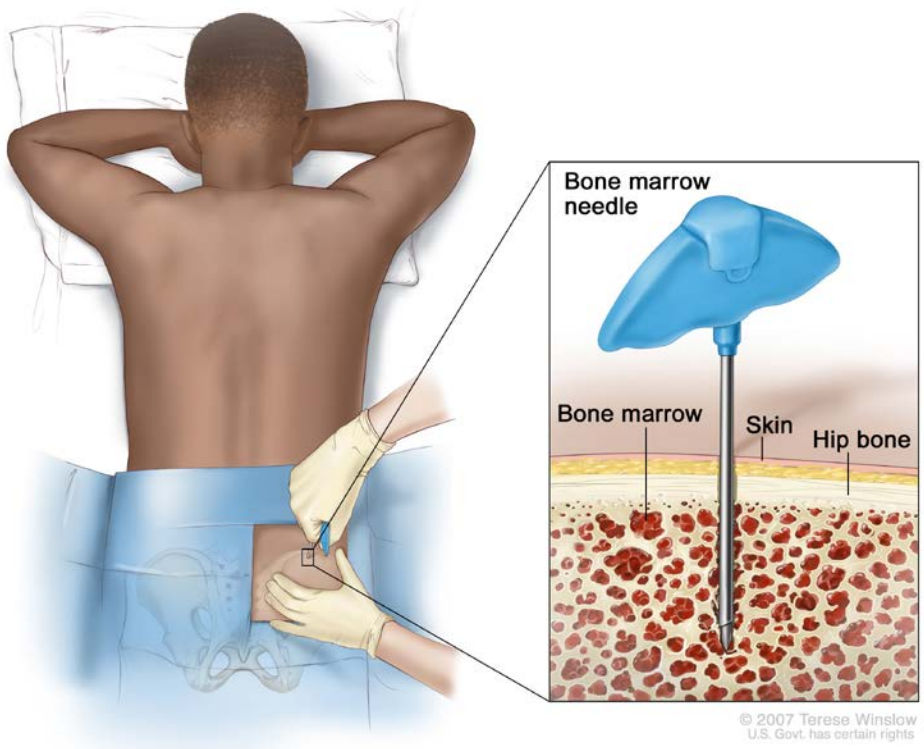
- Bone marrow aspirate
- Bone marrow biopsy

Your bone marrow is like a sponge holding liquid and cells. An aspirate takes some of the liquid and cells out of the sponge, and a biopsy takes a piece of the sponge.

The samples are usually taken from the back of the hip bone (pelvis). You'll likely lie on your belly or side. Your care team will first clean the area, sedate you, and numb your skin and the outer surface of your bone.

Bone marrow aspirate and biopsy

Samples of bone and liquid marrow are removed in a biopsy. A needle is inserted into the bone to remove these samples.



For an aspirate, a hollow needle will be pushed through your skin and into the bone. Liquid bone marrow will then be drawn into a syringe. For a biopsy, a wider needle will be used to remove a core sample. You may feel bone pain at your hip for a few days afterward. Your skin may bruise.

Genetic cancer risk testing

We inherit our genes from our parents. In hereditary or genetic syndromes, your siblings, your parents' siblings, and your grandparents often have the same change or mutation.

Certain genetic syndromes increase the risk of developing soft tissue sarcoma. Mutations in different genes cause each of these genetic syndromes.

While these cases of inherited genetic syndromes are rare, testing for gene mutations that cause these syndromes may, in the future, help treat bone cancer and monitor you or your family members for other cancers.

Share what you know about your personal health and family history with your care team. If your health care provider thinks you may have a genetic syndrome that is causing your cancer, you may benefit from genetic testing.

Testing should be carried out by a pathologist who’s experienced in genetic testing techniques. A genetic counselor may speak with you about the results. A genetic counselor is an expert who has special training in genetic diseases.

Biomarker testing

Biomarker testing is performed on a case-by-case basis and may increase the time it takes for biopsy results. Biomarker testing is sometimes called molecular testing or tumor profiling, tumor sequencing, gene expression profiling, or genomic testing.

Your care team will recommend the best types of biomarker testing for you.

Ewing sarcoma

A cell must make a copy of its chromosomes before dividing into 2 cells. Sometimes, there are mistakes in the copies.

One type of mistake is when parts of 2 chromosomes break off and switch with each other. This is called a translocation. It can result in a fusion of 2genes. A translocation between chromosomes 11 and 22 is common in Ewing sarcoma. This is represented as t(11;22).

Ewing sarcoma is also associated with the fusion of the *EWS* gene *(EWSR1)* on chromosome 22 with various members of the *ETS* gene family from chromosome 11, with *FLI1* being the most common.

Almost everyone with Ewing sarcoma will have one of the possible gene fusions. This is not inherited from your birth parents but results from an error of cell division while your tissues were growing. This is called a somatic mutation or somatic change.

Tumor mutation testing

Tumor mutation testing uses a sample of your tumor or blood to see if the cancer cells have any specific DNA mutations. Only the tumor is tested, not the rest of your body. This is a different type of DNA testing than the genetic testing for mutations you may have inherited from your birth parents.

Examples of tumor mutation testing include next-generation sequencing and polymerase chain reaction (PCR).

Key points

- A diagnosis of bone cancer is confirmed using a biopsy. It's recommended that the biopsy be performed by an experienced surgeon at the treatment center familiar with your diagnosis.
- A sample from a biopsy of your tumor might be tested to look for biomarkers.
- Your health care provider might refer you for genetic testing to learn more about your cancer.

Questions to ask

- What type of bone cancer do I have? What does this mean in terms of my prognosis and treatment options?
- From the tests that I have had, is my cancer localized to the bone or has it spread to other parts of my body?
- Is there a sarcoma cancer center or hospital nearby that specializes in this type of cancer?
- What tests will I have? How often will they be repeated?
- Will my insurance pay for these tests? Where can I find help to pay for tests?
- Who will talk with me about the next steps and when?

What's next?

After all of these tests, your doctor will talk to you and explain your complete diagnosis. You may want to ask another doctor to review your test results and treatment plan. This is called getting a second opinion.

The next chapter provides an overview of the many different treatment options for bone sarcoma.

3
Types of treatment

16 Care team

17 Surgery

18 Physical and occupational therapy

18 Fertility counseling

19 Drug therapy

20 Radiation therapy

21 Destroying tumors directly

22 Clinical trials

23 Supportive care

24 Survivorship

25 Key points

25 Questions to ask

Treatment for bone cancer is based on the type, size, and location of the tumor. This chapter presents a general overview of different types of treatment and what to expect.

Care team

Treating bone cancer takes a team approach. Treatment decisions should involve a multidisciplinary team of professionals with knowledge and experience in bone cancer. This team is united in planning and implementing ~~of~~ your treatment. Ask who will coordinate your care.

Some members of your care team will be with you throughout cancer treatment, while others will be there for parts of it. Get to know your care team and help them get to know you.

Your care team should include the following core group:

- **An orthopedic oncologist or orthopedic surgeon** specializes in the diagnosis and treatment of bone and soft tissue tumors. They treat all areas of the body except the neck and skull and inside of the chest and abdomen. If your tumor is in one of these areas, a different type of surgeon may be involved in your care.
- **A medical or pediatric oncologist** treats cancer using systemic drug therapy.
- **A radiation oncologist** prescribes and plans radiation therapy to treat cancer.
- **A musculoskeletal radiologist** interprets the results of x-rays and other imaging tests.
- **A bone pathologist** interprets the cells and tissues removed during a biopsy or surgery.

Depending on your care, your team might also include the following specialists:

- **A thoracic surgeon** performs operations in the chest.
- **A plastic surgeon** performs operations to improve function or reconstruct missing areas.
- **An interventional radiologist** performs needle biopsies, ablation therapies, and embolization. They also implant ports for drug therapy.
- **A physiatrist** is a medical health care professional who specializes in physical movement and rehabilitation.
- **A vascular surgeon** specializes in procedures involving arteries, veins, and lymph circulation.
- **A surgical oncologist or general surgeon** has specialized knowledge and experience related to the diagnosis and preoperative, operative, and postoperative care of the whole person.
- **A neurosurgeon** specializes in surgery of the nervous system, especially the brain and spinal cord.
- **An orthopedic spine surgeon** specializes in the diagnosis and treatment of spinal diseases and conditions.

Surgery

Treating most bone sarcomas starts with surgery. Surgery is an operation or procedure to remove cancer from the body.

The type of surgery depends on the size, location, and number of tumor(s), and should be considered based on your lifestyle and needs.

Surgery can be used to:

- Remove the cancer at the main site. Sometimes, surgery can remove areas of cancer that are spreading.
- Relieve pain or discomfort.

The primary goal of surgery is to remove the cancer. Your surgeon must cut out the tumor with a rim of healthy tissue around the edge. This is called a surgical margin.

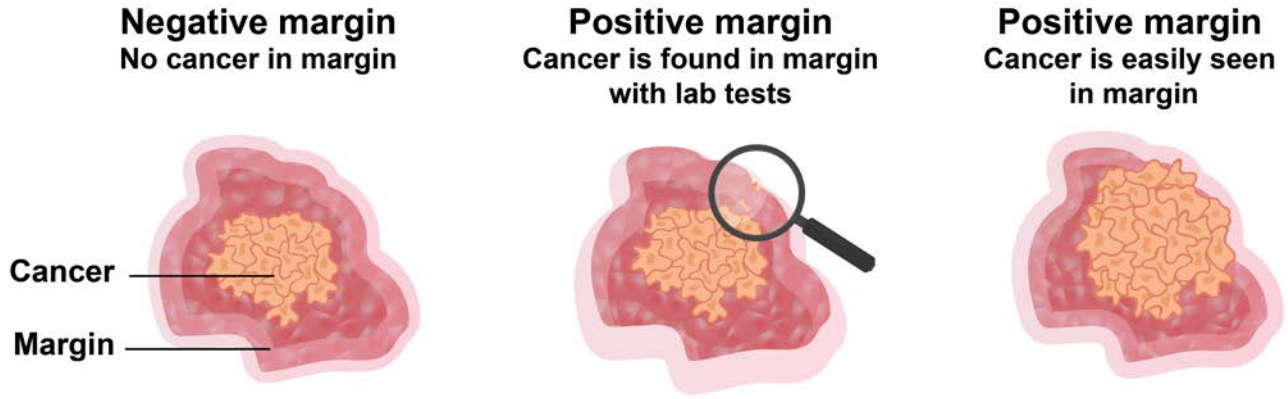
A clear, or negative, margin resection is when no cancerous cells are found in the tissue around the edge of the tumor. This means the margin is negative for cancer.

An involved, or positive, margin indicates that there is microscopic evidence of tumor cells at the edge of the resected tumor. In this case, it is assumed that cancer cells were therefore left in the body, at the site of the positive margin. This means that the margin is positive for cancer.

A surgeon who's an expert in your type of bone cancer should be consulted before deciding upon surgical treatment options. Not all surgeons or cancer centers are experienced in the types of surgery described below.

Surgical margin

The tumor will be removed, along with some normal-looking tissue around its rim. The normal-looking tissue is called the surgical margin. The surgical margin will be tested for cancer. Postoperative therapy is based on if there is cancer in the margins.



The type of surgery is based on many factors, including your wishes and the best choice for maintaining function or quality of life.

Limb-sparing surgery

For bone cancer in the limbs, the goal of surgery, whenever possible, is to preserve or maintain limb function. This is called limb-sparing surgery or limb salvage surgery. Reconstruction of the limb after limb salvage surgery depends on the specific resection needed to remove the bone cancer, and may include metal or bone replacement options.

Rotationplasty

In a rotationplasty, the bottom of the thigh bone, knee, and top of the shin bone (tibia) are removed. The lower leg is then rotated and attached to the femur.

The ankle joint acts as a knee joint. The intention of this surgery is to recreate knee function when it otherwise would not be possible. A patient who has had rotationplasty uses a specially made prosthesis for walking, functioning in a way similar to a person who has had a below-knee amputation. Not all surgeons or cancer centers offer this type of surgery.

Amputation

Amputation is surgery to remove part or all of a limb, such as an arm or a leg. It is done less frequently than limb-sparing surgery or rotationplasty for bone sarcomas. There are different types of amputations. Some types include reconstructive surgery, bone grafts, or implanted pieces.

Seek a peer support group to connect with others who have lost a limb. This will aid in your recovery and help you to maintain an active lifestyle.

Palliative surgery

Surgery to ease pain or discomfort is called palliative surgery. It might be used to remove tumors that are causing pain.

Palliative surgery is sometimes offered to fix a broken bone or help prevent a cancerous bone from breaking.

Physical and occupational therapy

Physical and occupational therapy will be part of any limb-sparing surgery, rotationplasty, amputation, or reconstructive surgery. Physical therapy helps your body move and function. Occupational therapy helps with daily life skills.

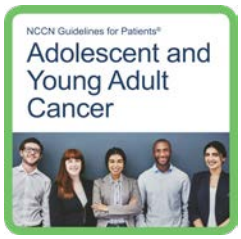
Fertility counseling

Some types of treatment can affect your fertility, the ability to have children. If you think you want children in the future, ask your care team how cancer and cancer treatment might change your fertility. To preserve your fertility, you may need to take action before starting cancer treatment. Those who want to have children in the future should be referred to a fertility specialist to discuss the options before starting treatment.

The goal of fertility preservation is to -keep your options open, whether you know you want to have children later in life or aren't sure now. Fertility and reproductive specialists can help

you sort through what may be best for your situation.

For more information on fertility preservation, see *NCCN Guidelines for Patients: Adolescent and Young Adult Cancer,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Changes in fertility

Treatment might cause your fertility to be temporarily or permanently impaired or interrupted. Loss of fertility is related to your age at the time of diagnosis, treatment type(s), treatment dose, and treatment length. Talk to your care team about your concerns and if you are planning a pregnancy.

Preventing pregnancy during treatment

Preventing pregnancy during treatment is important. Cancer and cancer treatment can affect the ovaries and damage sperm. So, becoming pregnant or having one's partner become pregnant during treatment should be avoided.

People who are capable of giving birth should take a pregnancy test before treatment begins.

Drug therapy

Drug therapy works throughout the body. It includes chemotherapy, targeted therapy, and immunotherapy.

Drug therapy might be used alone or with other therapies. Goals of drug therapy should be discussed before starting treatment.

All treatments listed in this guide are recommended and appropriate. When helpful, NCCN experts also assign a level of preference to their recommendations for drug therapies:

- **Preferred therapies** have the most evidence they may work better and be safer than other therapies.
- **Other recommended therapies** can provide effective results but may have less evidence, more side effects, or may not work quite as well as preferred therapies.
- **Therapies used in certain cases** work best for individuals with specific cancer features or health circumstances.

Chemotherapy

Chemotherapy kills fast-dividing cells throughout the body, including cancer cells and normal cells. More than one chemotherapy drug may be used to treat your type of cancer.

Some chemotherapy drugs are liquids that are infused into a vein or injected under the skin with a needle. Other chemotherapy drugs may be given as a pill that is swallowed.

In most cases, chemotherapy is given in cycles of treatment days followed by days of rest.

This allows the body to recover before the next cycle. Cycles vary in length depending on which chemotherapy is used. You might spend time in the hospital during treatment.

Cytoprotective medication

If you're given certain types of chemotherapy (such as ifosfamide), you might also be prescribed a cytoprotective medication called mesna (Mesnex). This medication decreases the risk of certain side effects from chemotherapy, like bleeding in the bladder.

Targeted therapy

Targeted therapy is a form of systemic drug therapy that focuses on specific or unique features of cancer cells. These drugs stop the action of molecules that help cancer cells grow and/or survive.

Targeted therapies seek out how cancer cells grow, divide, and move in the body. Some targeted therapy drugs block the chemical signals that tell cancer cells to grow. Other targeted therapy drugs block signals that cause new blood vessels to form.

Immunotherapy

Immunotherapy is systemic drug therapy that increases the activity of your immune system. By doing so, it improves your body's ability to find and destroy cancer cells. Immunotherapy can be given alone or with other types of treatment.

Radiation therapy

Radiation therapy uses high-energy rays to kill cancer cells and shrink tumors. It's usually prescribed for several weeks at a time, once a day or possibly once a week depending on your situation.

Radiation therapy can be given alone or with certain systemic therapies.

"Don't let yourself stop doing the things you enjoy doing. Whatever your hobbies are, whatever things you like to do, keep doing them. It'll help you feel more like yourself and stay positive during what is an extremely exhausting, mentally straining time."



When used as supportive care to help ease pain or discomfort caused by cancer, it is called palliative radiation therapy.

Different types of radiation therapy are used to treat bone cancer. These allow for safer, higher doses of radiation.

Types of radiation therapy that may be used to treat your cancer include:

- High-energy radiation beams over several treatments
- One precise, large dose of radiation
- Computer software and CT images to aim beams that match the shape of the tumor
- Small beams of different strengths to match the shape of the tumor
- Protons, carbon ions, or other heavy ions
- A solid implant placed into the body, similar to beads or seeds
- Radiopharmaceuticals, such as radium-223 (Xofigo), that contain a radioactive substance

Destroying tumors directly

The treatments in this section treat tumors by inserting a probe or special medical beads directly into the tumor.

Ablation

Ablation uses extreme cold or extreme heat to destroy cancer cells. It can destroy small tumors with little harm to nearby tissue.

Ablation uses a special needle, called a probe, which is inserted into the tumor directly. The probe may be guided into place with a CT scan, ultrasound, or other imaging tests.

In cryotherapy ablation, a gas is passed through the probe to freeze it, like treating a mole or wart. In radiofrequency ablation, the probe emits radio waves to heat the tumor and destroy it.

Embolization

Embolization treats tumors by cutting off their blood supply. To do this, a catheter is inserted into an artery using a needle and guided to the tumor. Once in place, beads are inserted into the tumor to block the blood flow.

In chemoembolization, the beads are coated with chemotherapy. In radioembolization, the beads are radioactive.

“Your medical team is your greatest asset and friend(s), especially the nurses. They are all highly skilled and sincerely dedicated to help you however they can."

Clinical trials

A clinical trial is a type of medical research study. After being developed and tested in a lab, potential new ways of fighting cancer need to be studied in people.

If found to be safe and effective in a clinical trial, a drug, device, or treatment approach may be approved by the U.S. FDA.

Everyone with cancer should carefully consider all of the treatment options available for their cancer type, including standard treatments and clinical trials. Talk to your health care professional about whether a clinical trial may make sense for you.

Phases

Most cancer clinical trials focus on treatment and are done in phases.

- **Phase 1** trials study the safety and side effects of an investigational drug or treatment approach.
- **Phase 2** trials study how well the drug or approach works against a specific type of cancer.
- **Phase 3** trials test the drug or approach against a standard treatment. If the results are good, it may be approved by the FDA.
- **Phase 4** trials study the safety and benefit of an FDA-approved treatment.

Who can enroll?

It depends on the clinical trial's rules, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, or general health. They ensure that participants are alike in specific ways and that the trial is as safe as possible for the participants.



Finding a clinical trial

In the United States

NCCN Cancer Centers
NCCN.org/cancercenters

The National Cancer Institute (NCI)
cancer.gov/about-cancer/treatment/clinical-trials/search

Worldwide

The U.S. National Library of Medicine (NLM)
clinicaltrials.gov

Need help finding a clinical trial?

NCI's Cancer Information Service (CIS)
1.800.4.CANCER (1.800.422.6237)
cancer.gov/contact

Informed consent

Clinical trials are managed by a research team. This group of experts will review the study with you in detail, including its purpose and the risks and benefits of joining. All of this information is also provided in an informed consent form. Read the form carefully and ask questions before signing it. Take time to

discuss it with people you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

Will I get a placebo?

Placebos (inactive versions of real medicines) are almost never used alone in cancer clinical trials. It's common to receive either a placebo with a standard treatment, or a new drug with a standard treatment. You'll be informed, verbally and in writing, if a placebo is part of a clinical trial before you enroll.

Are clinical trials free?

There's no fee to enroll in a clinical trial. The study sponsor pays for research-related costs, including the study drug. But you may need to pay for other services, like transportation or childcare, due to extra appointments. During the trial, you'll continue to receive standard cancer care. This care is often covered by insurance.

Supportive care

Supportive care is an important part of cancer care. The goal is to improve your quality of life during and after cancer treatment. Supportive care is for everyone with cancer and their families, not just for those at the end of life. It's also known as palliative care.

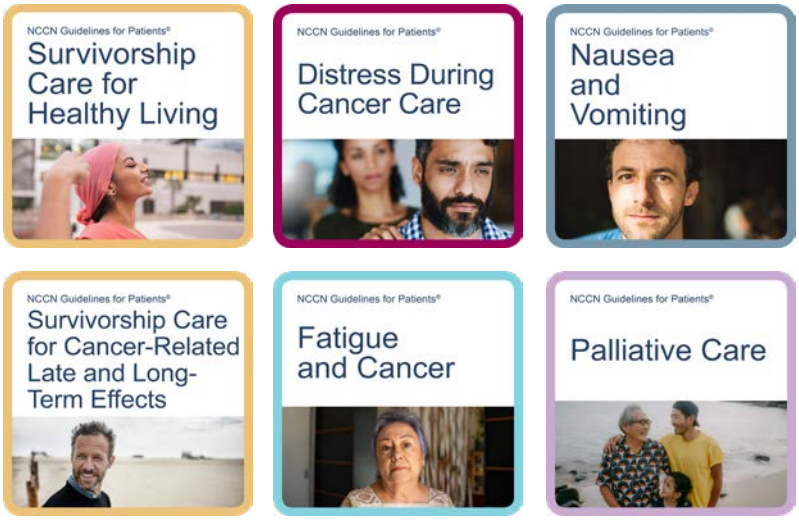
Supportive care includes a wide range of services. Supportive care prevents or manages the symptoms of cancer and the side effects of cancer treatment, like pain and cancer-related fatigue. It also addresses the mental, social, emotional, and spiritual concerns faced by people with cancer.

Supportive care provides help with additional needs, such as:

- Making treatment decisions
- Coordinating your care
- Planning for advance care and end of life
- Paying for care

Supportive care resources

More information on supportive care is available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Read more about the types of support you may receive in *NCCN Guidelines for Patients: Palliative Care*, available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Side effects

All cancer treatments can cause unwanted health issues called side effects. Side effects depend on many factors. These factors include the drug type and dose, length of treatment, and the person.

Treatment can cause several side effects. Some side effects may be harmful to your health. Others may just be unpleasant. A few are very serious.

You'll be monitored closely for side effects. There are also ways to prevent some side effects.

Ask for a complete list of side effects of your treatments. Also, tell your treatment team about any new or worsening symptoms. There may be ways to help you feel better.

Late effects

Late effects are side effects that happen months or years after you're told you have cancer or after the treatment is over. Late effects may be caused by cancer or cancer treatment. They may include physical, mental, and personal life issues. They could also include second cancers.

The sooner late effects are treated, the better. Ask your care team about what late effects could occur. This will help you know what to look for.

Survivorship

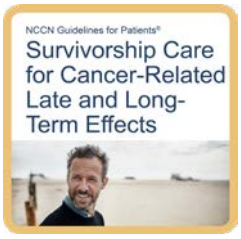
A person is a cancer survivor from the time of diagnosis until the end of life. After treatment, your health will be monitored for side effects of treatment and the return of cancer. This is part of your survivorship care plan.

Try your best to keep any follow-up health care visits and imaging test appointments. Seek good routine medical care, including regular visits for preventive care and cancer screening.

A personalized survivorship care plan will contain a summary of possible long-term effects of treatment and list follow-up tests. Find out how your primary care provider will coordinate with specialists for your follow-up care.

Read more about how to live healthily as a survivor in *NCCN Guidelines for Patients: Survivorship Care for Healthy Living* or in *NCCN Guidelines for Patients: Survivorship Care for Late and Long-Term Effects*, available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.





Key points

- Surgery is an operation or procedure to remove cancer from the body. The type of surgery depends on the size, location, and amount of tumors.
- Systemic drug therapy works throughout the body. It includes chemotherapy, targeted therapy, and immunotherapy.
- Radiation therapy uses high-energy x-rays, protons, gamma rays, and other sources to kill cancer cells and shrink tumors.
- Ablation uses extreme cold or extreme heat to destroy cancer cells.
- Embolization treats tumors by cutting off their blood supply.
- Supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life.

Questions to ask

- Can you recommend a health care professional, hospital, or cancer center that specializes in my type of bone cancer?
- Which treatment(s) do you recommend and why?
- What can I expect from treatment?
- Am I candidate for a clinical trial?
- How will you treat side effects?

4
Osteosarcoma

28 Drug therapy

29 Low-grade osteosarcoma

29 High-grade osteosarcoma

30 Metastatic disease at diagnosis

30 Surveillance

30 If cancer comes back

31 Key points

31 Questions to ask

Osteosarcoma is the most common type of bone cancer. Most often, it usually occurs around the knee or shoulder. Treatment is a combination of therapies, usually chemotherapy and surgery. Together, you and your care team will choose a treatment plan that is best for you.

Osteosarcoma is cancer that starts in bone-forming cells called osteoblasts. The cause is not known. Osteosarcoma can occur in any bone, but it's commonly found in large bones. It develops in the area of bone with the fastest growth rate.

Osteosarcoma tends to occur in bones of the:

- Shin (near the knee)
- Thigh (near the knee)
- Upper arm (near the shoulder)
- Hip bone (pelvis)

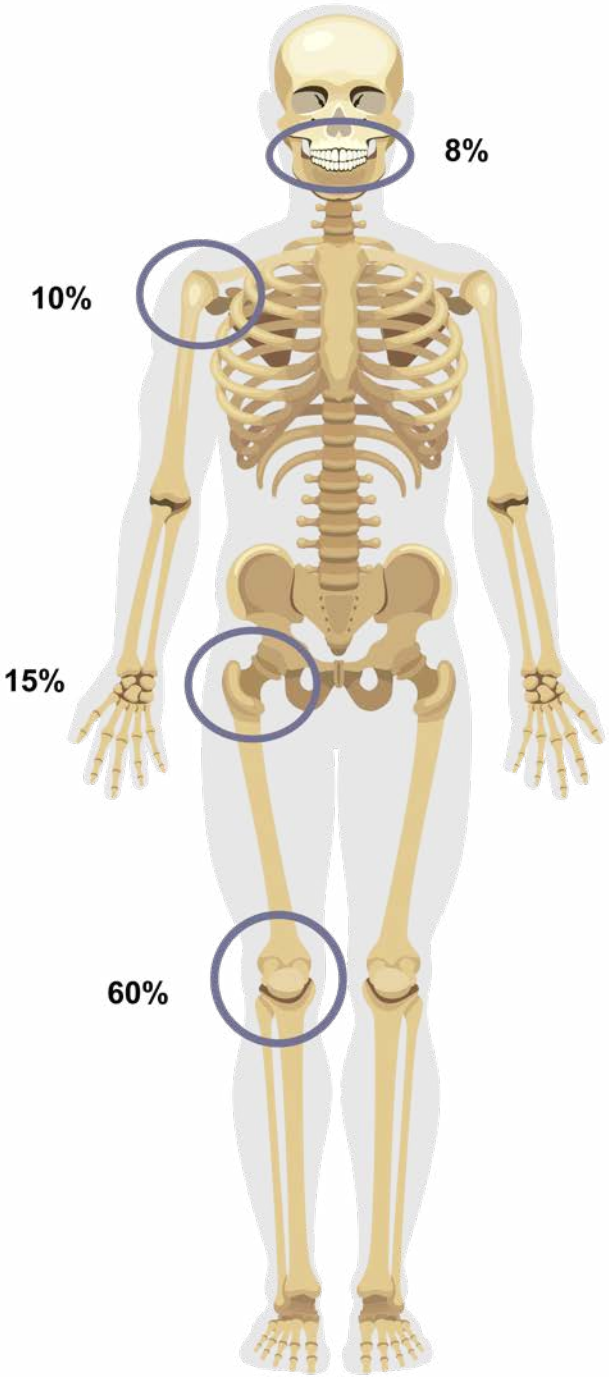
There are 2 main types of osteosarcoma:

- Low-grade osteosarcoma (growing on the bone surface)
- High-grade osteosarcoma (typically growing around the knee)

Sometimes, osteosarcomas can grow outside of the bone, in the muscles or cartilage.

These may be treated as soft tissue sarcomas, usually with ifosfamide or platinum-based therapy (cisplatin/doxorubicin). For more information, see *NCCN Guidelines for Patients: Soft Tissue Sarcoma*, available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.

Common locations where osteosarcoma is found. These tend to be areas that grow fastest.



Drug therapy

Chemotherapy and other drug therapies are part of treatment for osteosarcoma. First-line chemotherapies are those given first. Second-line drug therapy options are used when disease progresses, does not respond to first-line options, or relapses. See **Guide 2** and **Guide 3**.

"Cancer can temporarily rob you of your energy, but it can't steal your burning desire to get it all back."

Guide 2
Osteosarcoma: Initial drug therapy options

Preferred options	• Cisplatin, -Doxorubicin • High-Dose Methotrexate, Cisplatin, Doxorubicin, Leucovorin
Other recommended options	• Cisplatin, Doxorubicin, Ifosfamide, Mesna, High-Dose Methotrexate, Leucovorin

Guide 3
Osteosarcoma: Second-line drug therapy options

Preferred options	• High-Dose Ifosfamide, Mesna, with or without Etoposide • Regorafenib • Sorafenib
Other recommended options	• Cabozantinib • Cyclophosphamide, Topotecan • Gemcitabine with or without Docetaxel • Everolimus, Sorafenib
Used in some cases	• Cyclophosphamide, Etoposide, Mesna • Carboplatin, Etoposide, Ifosfamide, Mesna • High-Dose Methotrexate • Etoposide, High-Dose Methotrexate, Ifosfamide, Mesna • Radium-223 for relapsed or refractory disease beyond second-line therapy

Low-grade osteosarcoma

Treatment for low-grade osteosarcoma is surgery to remove the tumor, usually without chemotherapy. Drug therapy might follow surgery if a part of the tumor is a higher-grade than expected.

Osteosarcoma that grows on the outside of the bone usually only requires surgery.

However, certain types of external bone osteosarcomas, where the cells have become more primitive, are not considered to be low-grade tumors and should be treated as high-grade.

Osteosarcoma on the surface of the bone

Periosteal osteosarcoma starts on the surface of the bone. Treatment is surgery to remove the tumor with no cancer remaining in the body (negative margins). Systemic drug therapy might be given before surgery to reduce the size of tumor or the amount of cancer.

High-grade osteosarcoma

Drug therapy

Treatment for high-grade osteosarcoma usually starts with chemotherapy. The goal of chemotherapy is to kill as many of the cancer cells as possible before surgery.

After chemotherapy, you'll have imaging tests to check your cancer and assess how well the chemotherapy worked, then plan surgery. For preoperative chemotherapy options, see **Guide 2.**

Diagnosis vs. Prognosis

What's the difference between your diagnosis and your prognosis? These 2 words sound alike but they're very different.

Diagnosis: The identification of an illness based on tests. The diagnosis names what illness you have.

Prognosis: The likely course and outcome of a disease. The prognosis predicts how your disease will turn out.

You might receive chemotherapy after surgery, depending on the outcome of your surgery. This may depend on if there are still tumors or cancer cells in your body after surgery. Most younger people receive additional chemotherapy after surgery.

Surgery or local control therapy

High-grade osteosarcomas are also treated with surgery to remove all of the cancer that can be seen in the body with MRI or CT scans. Imaging scans beforehand help determine if the osteosarcoma can be removed with surgery.

Most osteosarcomas are removable with surgery, with the goal being a **negative** margin, or cutting out as much of the tumor as safely as possible. This could require limb-sparing surgery or amputation.

Rarely, the tumor can't be removed by surgery. In this case, treatment involves radiation therapy or systemic therapy.

Metastatic disease at diagnosis

Sometimes more cancer growths are found in the internal organs in addition to the bone. These are often removable and are called metastatic growths.

Metastatic growths might be treated with surgery (if possible); drug therapy; radiation therapy; or other therapies, such as ablation. Before drug therapy is started, biomarker testing might be done to assess if there are any molecular targets that a specific drug could act upon.

Surveillance

After completing treatment, you'll be monitored on a regular basis to watch for signs that cancer has returned, called surveillance.

Surveillance includes a physical exam, blood and imaging tests, functional movement tests, and regular visits with your orthopedic surgeon and oncologist.

If cancer comes back

If cancer returns, treatment is a new drug therapy and/or a new surgery. Surgery is not always an option. Systemic drug therapy options can be found in **Guide 3.**

"Be your own advocate. Talk to someone who has gone through the same thing as you. Ask a lot of questions, even the ones you are afraid to ask. You have to protect yourself and ensure you make the best decisions for you, and get the best care for your particular situation."



Treatments for relapse and disease progression include:

- Surgery to remove the tumor, if possible
- Clinical trial
- Radiation therapy (may include radiopharmaceuticals)
- Embolization/ablation (see *Destroying tumors directly* in Chapter 3)
- Best supportive care

After treatment for relapse, you'll have imaging tests. If your cancer responds to treatment, then you'll be monitored for the return of cancer or to see if it gets worse.

Key points

- Osteosarcoma is the most common type of bone cancer. It often grows around the knee or shoulder.
- Treatment most commonly involves drug therapy and surgery. Some osteosarcomas only require surgery.
- Osteosarcomas can metastasize in the lungs, bone, or internal organs. These are sometimes removable with surgery.
- After completing treatment, you'll be monitored to watch for signs that cancer has returned. This is called surveillance.

Questions to ask

- Which treatment option is proven to work best for my type of cancer, age, overall health, and other factors?
- What are the available surgery options for my type of cancer?
- Where can I find more information on the types of surgery available to me?
- Can you recommend an experienced surgeon or where I can go for a second opinion?
- What are the pros and cons of the different surgery options? Who can help me decide?

5
Ewing sarcoma

34 Initial treatment

34 Checking your cancer

36 Metastatic disease

37 Key points

37 Questions to ask

Ewing sarcoma is a group of cancers that start in the bone or soft tissue. Treatment usually begins with drug therapy.

Ewing sarcoma is a cancerous tumor that occurs in bone, muscle, organs, or nerves.

There are 2 types of Ewing sarcoma:

- Ewing sarcoma of bone
- Ewing sarcoma of soft tissues (muscles, organs, or nerves)

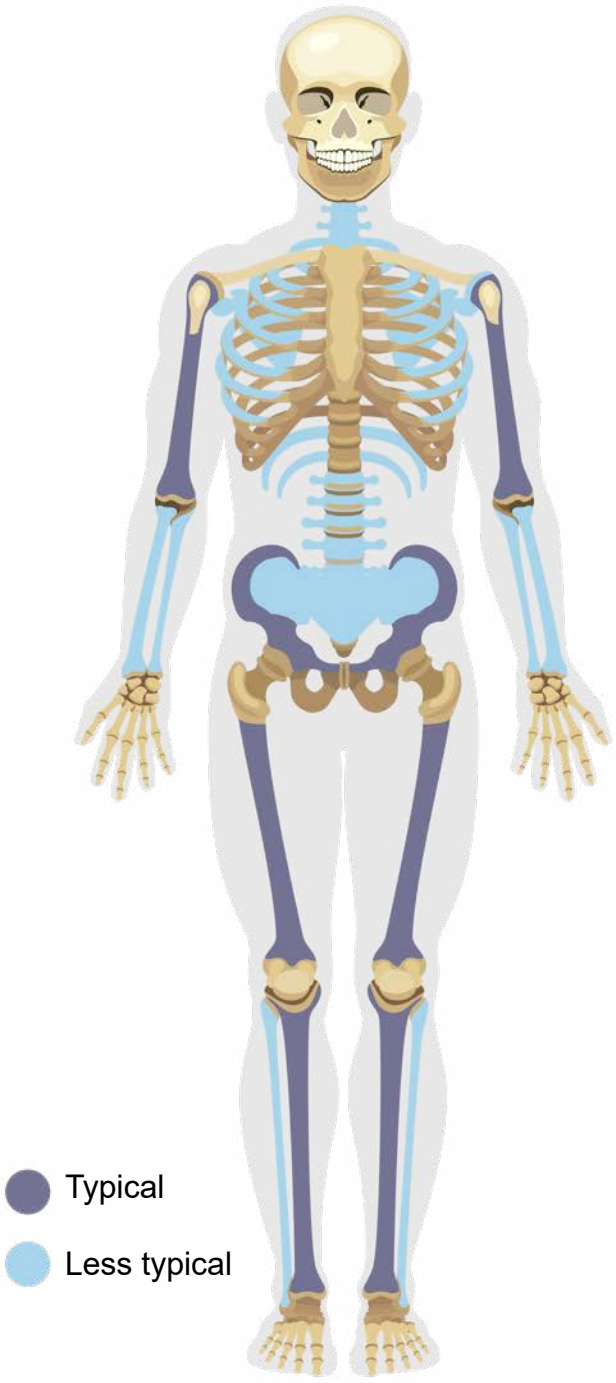
Ewing sarcoma of bone usually occurs in the thigh bone, hip bone, and bones of the chest. However, any bone may be affected. When arising in a long bone, the long center shaft is the most common site.

Ewing sarcoma can spread to other parts of the body, usually to the lungs, other bones, or bone marrow.

Ewing sarcoma of soft tissues is a soft tissue sarcoma. For more information, see *NCCN Guidelines for Patients: Soft Tissue Sarcoma,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Ewing sarcoma emerges in the darker-colored areas of bone more frequently than the lighter-colored areas.



Initial treatment

Chemotherapy is often the initial, or first, treatment for all types of Ewing sarcoma. It includes a combination of chemotherapies. This is called multi-agent chemotherapy. You'll have at least 9 weeks of this chemotherapy.

For initial drug therapy options, see **Guide 4.**

Checking your cancer

After first-line chemotherapy, your cancer will be rechecked. This will determine if the cancer is improved or not responding to treatment.

Possible tests to re-stage your cancer might include:

- Chest CT
- MRI with or without CT (both with contrast) of primary site
- X-rays of primary site
- FDG-PET/CT (head-to-toe) or bone scan

Stable or improved disease

If your cancer has improved with drug therapy or hasn't gotten worse (remained stable), then you'll receive a new treatment. This treatment focuses on the main tumor site.

This therapy might be:

- Surgery to remove the tumor (wide excision). Limb-sparing surgery or amputation might be necessary.
- Radiation therapy

The goal is to surgically remove all cancer or destroy all cancer cells with radiation therapy. Sometimes, surgery with radiation is recommended.

You'll likely need more rounds of systemic therapy after this round of treatment. The best treatment for you will be based on your cancer team's recommendations and your wishes. Most young people will receive additional systemic therapy after surgery or radiation.

Guide 4
Ewing sarcoma: Initial drug therapy options

Preferred option	• VDC-IE: Vincristine, Doxorubicin*, Cyclophosphamide alternating with Ifosfamide, Mesna, Etoposide
Other recommended	• VAIA: Vincristine, Doxorubicin*, Ifosfamide, Mesna, Dactinomycin • VIDE: Vincristine, Ifosfamide, Mesna, Doxorubicin*, Etoposide

*Note: Dactinomycin may be substituted for Doxorubicin.

Not responding to treatment or disease progression

When cancer does not respond or progresses after primary treatment, radiation therapy and/or surgery might be considered to prevent the spread of cancer or to ease symptoms.

If cancer continues to progress or has metastasized, then options include a second-line systemic therapy or best supportive care.

For second-line systemic therapy options, see **Guide 5.**

Monitoring

After completing treatment, you'll be monitored for signs that cancer has returned. Cancer that returns is called relapse or recurrence. Monitoring includes physical exams, blood tests, and imaging scans.

If Ewing sarcoma relapses, treatment is systemic therapy with the possibility of radiation therapy or more surgery.

Guide 5
Ewing sarcoma: Treatment options for relapsed, progressed, or metastatic cancer

Preferred options	• Cyclophosphamide, Topotecan. Vincristine might be added. • Irinotecan, Temozolomide. Vincristine might be added.
Other recommended	• Cabozantinib • Docetaxel, Gemcitabine. Vincristine might be added. • High-Dose Ifosfamide, Mesna • Regorafenib
Used in some cases	• Ifosfamide, Mesna, Carboplatin, Etoposide. Vincristine might be added. • Lurbinectedin

Metastatic disease

Metastatic Ewing sarcoma is cancer that's found in more than one location. It may be cancer that has spread from the original, or first, location to other parts of the body.

Treatment for metastatic disease at diagnosis is different from treatment for cancer that has recurred or progressed. Treatment of cancer that has spread or metastasized is explained next.

Main tumor therapy

This therapy focuses on the main tumor after drug therapy. Options include surgery, radiation therapy, systemic drug therapy, or a combination of these treatments.

Metastasis might be treated separately from the main tumor. Lungs, bone, and bone marrow are the most common sites of cancer spread, or metastasis. Often, the lungs are easier to treat than cancer that has spread to other places. Treatment for lung metastasis might include surgery and/or radiation therapy.

Metastatic disease at diagnosis

Systemic therapy is the first treatment for all types of Ewing sarcoma, including metastatic disease as the initial diagnosis. Systemic therapy includes a combination of chemotherapies. Systemic therapy options for an initial diagnosis of metastatic Ewing sarcoma can be found in **Guide 6.**

Widely metastatic cancer

Widely metastatic cancer is where the cancer has spread to many different places throughout the body. Treatment for widely metastatic disease is either drug therapy with palliative surgery or palliative radiation to treat areas causing pain or discomfort. Other techniques might be used to treat metastases.

Biomarker testing might be done before continuing drug therapy.

Guide 6
Ewing sarcoma: Drug therapy options for metastatic disease at initial diagnosis

Preferred options	• Vincristine, Doxorubicin*, and Cyclophosphamide alternating with Mesna-Ifosfamide, Mesna, and Etoposide • Vincristine, Doxorubicin, Ifosfamide, Mesna, and Dactinomycin • Vincristine, Ifosfamide, Mesna, Doxorubicin*, and Etoposide • Vincristine, Doxorubicin*, and Cyclophosphamide

*Note: Dactinomycin may be substituted for Doxorubicin.

Key points

- Chemotherapy (a form of systemic drug therapy) is the first, or primary, treatment for Ewing sarcoma.
- Treatment after initial systemic drug therapy focuses on the main tumor site. The goal is to remove all cancer with surgery or destroy all cancer cells with radiation therapy.
- More systemic drug therapy might be given after surgery or radiation therapy.
- Treatment for an initial diagnosis of metastatic Ewing sarcoma is different from treatment for cancer that has spread during or after treatment.
- Metastatic disease at diagnosis is treated with a combination of chemotherapy drugs.

Questions to ask

- Which treatments do you recommend and why?
- How much will my insurance pay for treatment?
- Does this treatment offer a cure? If not, how well can treatment stop the cancer from growing?
- Does the order of treatments matter?
- What side effects can I expect from this treatment?

6
Chondrosarcoma

39 Treatment for primary tumors

40 Metastatic disease

41 If cancer comes back

42 Key points

42 Questions to ask

Chondrosarcoma starts in the cells that form cartilage. Treatment is usually surgery to remove the tumor. Together, you and your care team will choose a treatment plan for you.

Chondrosarcoma starts in the cells that form cartilage. It's most commonly found in the thigh bone, arm, or hip bone (pelvis).

Treatment for primary tumors

Treatment options are based on the type and location of the chondrosarcoma and your care team's recommendations. Remember, you have the power to decide what's best for you. Not treating the tumor is an option.

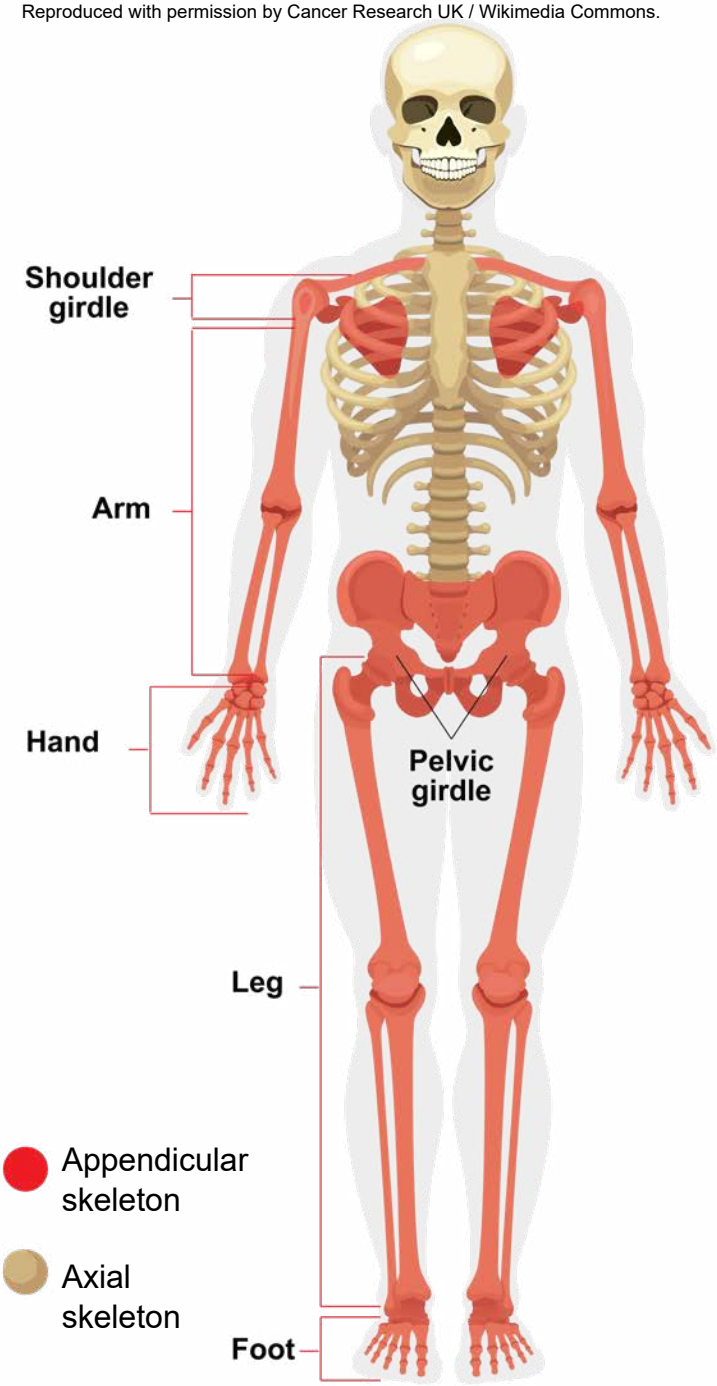
Atypical cartilaginous tumors

Atypical cartilaginous tumors are low-grade, typically benign tumors of the arms, shoulders, legs, and hip bones. Sometimes these tumors transform into a more aggressive cancer. If this happens, it would be treated more aggressively.

For chondrosarcoma, surgery to remove all of the tumor with a negative margin is the preferred treatment. Some tumors in the cartilage that are not chondrosarcoma may be treated with observation (watch and wait) or less extensive surgery.

Chondrosarcoma emerges in the appendicular skeleton. The appendicular skeleton includes the shoulder girdle, the upper limbs, the pelvic girdle, and the lower limbs. The axial skeleton includes the skull, rib cage, and spine.

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After completing treatment, you'll be monitored with the following tests for the return of the tumor:

- Physical exam
- X-rays every 6 to 12 months for 2 years, then yearly as needed
- Other imaging tests, such as CT with contrast, MRI, chest imaging, and bone scan, as needed

Malignant tumors

Malignant tumors are cancers that tend to spread. So, they need more aggressive treatment than benign tumors.

Treatment options include:

- If removable, the preferred treatment is surgery to remove all of the tumor. This might be limb-sparing or an amputation.
- If not removable or too close to important parts, radiation therapy might be given instead.

After treatment, your care team will check in with you regularly to make sure the cancer doesn't come back. See **Guide 7**.

Metastatic disease

Metastatic disease is cancer that has spread to other parts of the body. Treatment for metastatic chondrosarcoma is based on the amount of cancer that has spread.

Treatment focuses on reducing the number of metastases or amount of cancer.

A few metastases

If possible, you'll have surgery to remove all of the metastases.

Radiation therapy is an option for unremovable metastases. A clinical trial using systemic drug therapy is also an option.

Guide 7
Regular follow-up tests for chondrosarcoma

Physical exams
X-rays, CT scans, and MRI scans as needed
Chest scans every 3 to 6 months; this may include chest CT at least every 6 months for 5 years, then every year for at least 10 years
Checkups at every visit to assess how you're functioning at home

Widespread disease

Treatment options for widespread disease might include:

- Radiation therapy, surgery, and/or ablation therapy (see *Destroying tumors directly* in Chapter 3) for sites causing symptoms
- Targeted drug therapies, such as dasatinib (Sprycel) or pazopanib (Votrient)
- Clinical trial

Before starting drug therapy, you might get genetic biomarker testing. Tumors found with specific mutations may receive certain medicines. For tumors with *IDH1* mutations, for example, ivosidenib (Tibsovo) might be an option.

If cancer comes back

Cancer that has returned after a disease-free period is called a recurrence. When this is suspected, a biopsy is sometimes done to check.

If cancer returns at or near the same place, then the treatment options are:

- **Surgery** to remove all of the tumor, if possible. If surgery doesn't remove all of the cancer, then you might need more surgery or radiation therapy.
- **Radiation therapy** if the tumor is unremovable.



Warnings about supplements and drug interactions

You might be asked to stop taking or avoid certain herbal supplements when on a systemic therapy. Some supplements can affect the ability of a drug to do its job. This is called a drug interaction.

It is critical to speak with your care team about any supplements you may be taking. Some examples include:

- Turmeric
- Ginkgo biloba
- Green tea extract
- St. John's Wort
- Antioxidants

Certain medicines can also affect the ability of a drug to do its job. Antacids, heart or blood pressure medicines, and antidepressants are just some of the medicines that might interact with systemic therapy or supportive care medicines given during systemic therapy. Therefore, it is very important to tell your care team about any medicines, vitamins, over-the-counter (OTC) drugs, herbals, or supplements you are taking.

Bring a list with you to every visit.

Key points

- Chondrosarcoma starts in the cells that form cartilage.
- Treatment is based on the grade of the tumor and if it can be removed with surgery.
- Surgery to remove part of a tumor is limited to tumors that are thought to be non-cancerous. Surgery to remove all of a tumor is preferred.
- The primary treatment for non-metastatic chondrosarcoma is wide surgical resection.
- The goal of treatment for metastatic chondrosarcoma is to reduce the number of metastases or the amount of cancer in the body using surgery or radiation therapy.
- A clinical trial is an option for metastatic chondrosarcoma.

Questions to ask

- Is the tumor removable and how does this affect my treatment options?
- What side effects can I expect from treatment?
- What decisions must be made today?
- Who can help me decide which treatment is best for me?
- Is a clinical trial an option for me?

7
Chordoma

45 Treating chordomas

45 Monitoring

45 If cancer comes back

46 Key points

46 Questions to ask

7 Chordoma

Chordoma is a slow-growing cancer that usually starts in the lower spine or at the bottom of the skull, where it meets the spine. It can be difficult to treat because it often grows into the soft tissue around the bone.

Chordomas are a type of cancerous tumor that can occur anywhere along the spine, from the base of the skull to the tailbone. These tumors grow slowly, gradually invading nearby bone and soft tissue, like cartilage.

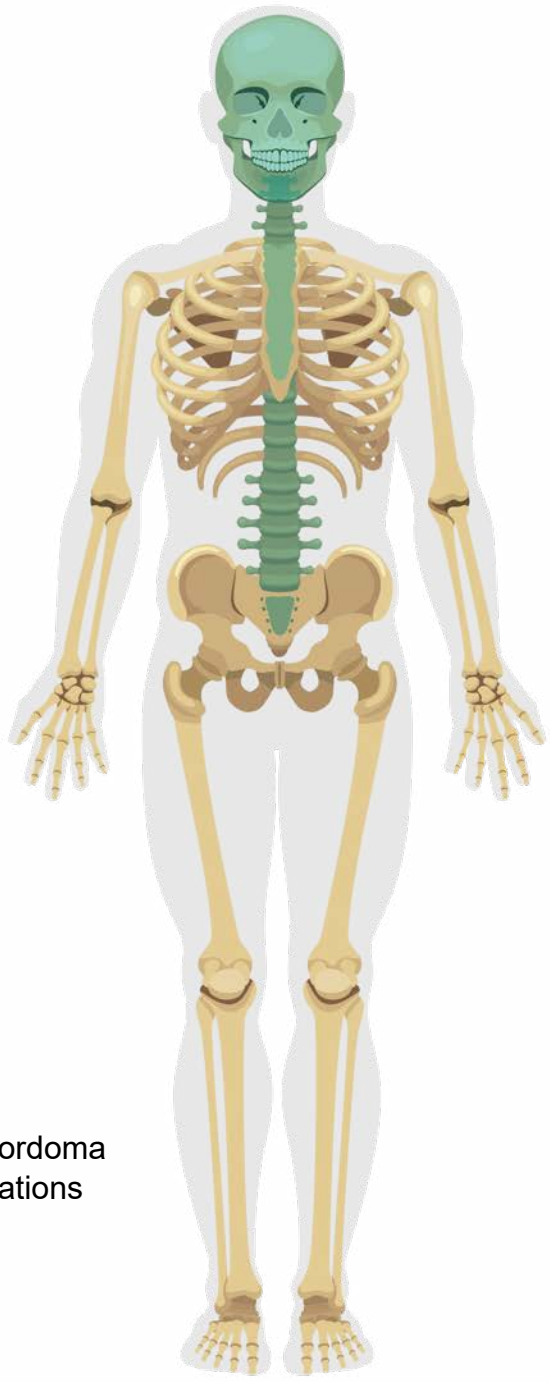
Tumors often involve critical structures, such as the brainstem, spinal cord, and important nerves and arteries. For this reason, chordomas are difficult to treat.

They can also come back after treatment—usually in the same place as the first tumor. This is called a local recurrence. In about 2 out of 5 people, the tumor eventually spreads, or metastasizes, to other parts of the body.

For some chordomas that have simpler cell structures, chemotherapies that are anthracycline- or gemcitabine-based are recommended. For more information, see *NCCN Guidelines for Patients: Soft Tissue Sarcoma,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Chordoma appears in the highlighted areas most frequently.



Chordoma locations

Treating chordomas

Chordomas are treated based on the location of the tumor:

- Tumors in the spine that include the lower back and tailbone (sacrococcygeal)
- Tumors at or in the bottom of the skull, where it meets the spine

A removable tumor is treated with surgery. The goal is to remove as much of the tumor as possible. Chordomas often grow outside of the bone into nearby vital organs, veins, and arteries. Parts of these tissues might also need to be removed and sewn back together.

An unremovable tumor might be treated with radiation therapy.

Tumors in the spine

For a removable tumor in the spine, surgery is the most important part of treatment. Radiation therapy might be given before, during, or after surgery.

For an unremovable tumor in the spine, radiation therapy without surgery might be an option.

Tumors in the skull base

For tumors at or in the base of the skull:

- If the tumor is removable, then surgery will be performed. Radiation therapy might be given before, during, or after surgery. A follow-up MRI with contrast will be done to see if radiation therapy or another surgery is needed.
- If the tumor is unremovable, then radiation therapy might be an option.

Monitoring

After completing treatment, you'll be monitored to watch for signs that cancer has returned. Monitoring includes imaging tests (x-rays, MRIs, or CTs) for up to 10 years or as needed. It also might include chest scans every 6 months and a chest CT every year.

If cancer comes back

When cancer returns, it's called a recurrence. In local recurrence, cancer returns close to where the initial (original) tumor was found. In metastatic recurrence, cancer is found in bones or other parts of the body far from the original site.

Treatment for either type of recurrence might include one or more of the following:

- Surgery
- Drug therapy
- Radiation therapy
- Ablation
- Clinical trial
- Best supportive care to relieve symptoms caused by cancer and improve quality of life

Treatments might be used alone or together. Most drug therapy options for chordoma recurrence are targeted therapies, such as imatinib (Gleevec), dasatinib (Phyrago, Sprycel), and sunitinib (Sutent). Other targeted therapies might also be given, such as pembrolizumab (Keytruda).

Key points

- Chordomas are treated based on the location of the tumor.
- The goal of surgery is to remove as much of the tumor as possible.
- An unremovable tumor might be treated with radiation therapy.
- When cancer returns, it's called a recurrence. In local recurrence, cancer returns close to where the initial tumor was found. In metastatic recurrence, cancer is found in bones or other parts of the body far from the original site.

Questions to ask

- Which treatment(s) do you recommend and why?
- Are there resources to help me pay for treatment or other care I may need?
- Does this treatment offer a cure? If not, how well can treatment stop the cancer from growing?
- Does the order of treatments matter?
- What side effects can I expect from this treatment?

8
Giant cell tumor of bone

49 Localized disease

49 Surveillance

49 Recurrence

49 Metastatic disease

51 Key points

51 Questions to ask

Giant cell tumor of bone (GCTB) is usually not cancer (benign). However, it is an aggressive, unpredictable disease that tends to return. Treatment is needed to prevent severe bone damage.

Giant cell tumor of bone (GCTB) is a rare, aggressive, non-cancerous tumor. It generally happens in adults between 20 and 40 years of age, when skeletal bone growth is complete. We don't know what causes a giant cell tumor.

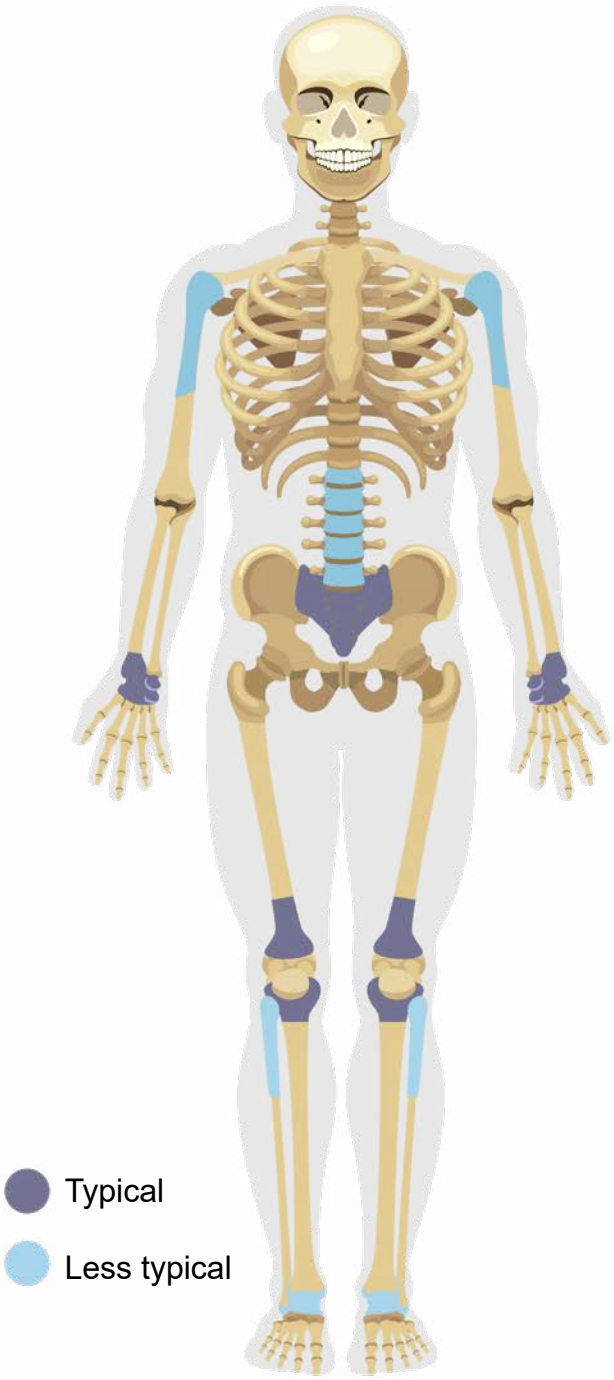
GCTB usually develops near a joint at the end of the bone. The knee is the most common location, but GCTB can be found in the bones of the arms and the legs. It can also affect flat bones, such as the breastbone or hip bone.

GCTB has a strong tendency to return to the same location. This is called local recurrence. Although rare, it may also reappear in the lungs.

When possible, treatment for a giant cell tumor almost always involves surgery to remove the tumor. Treatment aims to prevent damage to the bone near the affected joint.

Before starting treatment, you'll have a biopsy to confirm the diagnosis. You'll also have imaging tests to see if the tumor is in one area or has spread to distant sites.

Giant cell tumor of bone emerges in the darker-colored areas (the wrist, inner pelvis, and near the knees) most frequently.



Localized disease

In localized disease, the tumor is confined to one area. Treatment is based on whether the tumor is removable or unremovable as well as your overall health.

Removable tumor

A removable tumor is one that can be taken out with surgery. Frequently, the goal of surgery is to save normal bone and scrape out all of the tumor cells.

Sometimes, more surgery is required to remove all bone containing the tumor. This type of surgery can require reconstruction of the bone or joint.

At the time of surgery, your surgeon might use treatment like ablation or embolization to help prevent the return of the tumor.

Unremovable tumor

An unremovable tumor cannot be removed with surgery. This might be due to its location or other reasons, like if you have a serious health condition that prevents surgery.

Treatment options for an unremovable tumor include:

- Denosumab (preferred)
- Embolization (preferred)
- Radiation therapy

Denosumab (Xgeva) is given to prevent bone loss and fractures. This medicine stops the growth of the tumor and its destruction of the bone by blocking the signals from the tumor cells. The medicine does not, however, kill the tumor cells.

After treatment, you'll have imaging tests. If tests find that the tumor is now removable, you'll have surgery to remove the tumor. If the tumor cannot be removed with surgery, you'll begin surveillance.

Surveillance

Surveillance is when your care team monitors you for any signs that cancer has returned after you've completed treatment. Monitoring includes imaging tests of the surgical site. You'll also have chest imaging to look for lung metastases.

Recurrence

GCTB often returns to the same area, called recurrence.

There is also a concern it can spread to the lungs. You'll have chest imaging to see if there are any growths in the lungs.

If it's a local recurrence, denosumab might be given before surgery for a removable primary tumor.

Metastatic disease

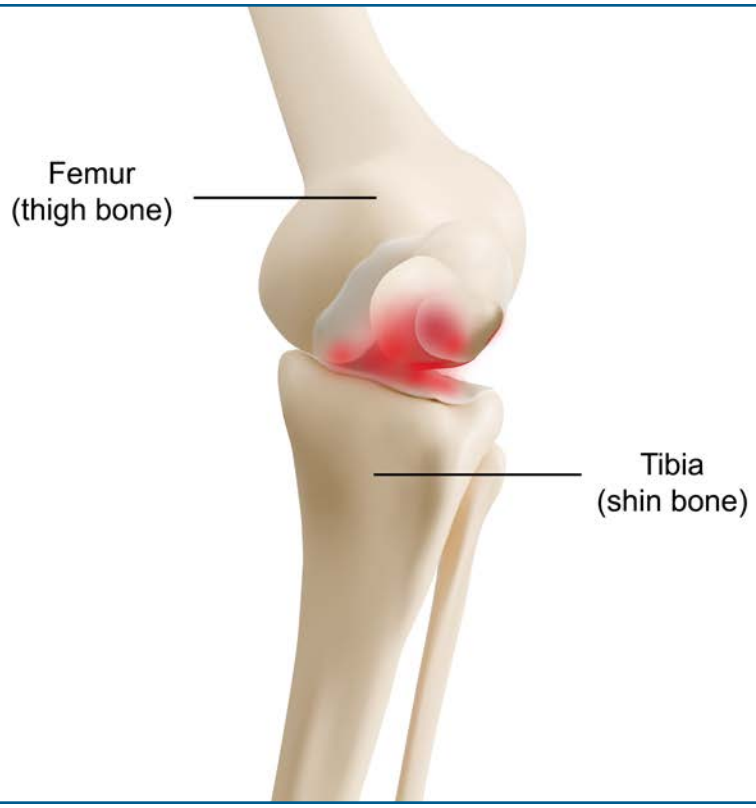
Metastatic disease at diagnosis is different from when GCTBs transform from an initial growth, because they are typically non-cancerous. But in some cases, GCTBs can transform into a more aggressive cancer.

If this happens, GTCBs are treated as osteosarcoma, which is explained in Chapter 4.

In metastatic disease, there is more than one tumor. While GCTB is usually benign, it can spread (metastasize) to the lungs or other bones.

- For removable tumors, surgery can target the main tumor and other growths.
- For unremovable tumors, treatment options include denosumab (preferred), radiation therapy, or observation. Observation is sometimes referred to as “watch and wait.” Ask what this might mean for you.

“Stay as active and eat as well as you can. Find a way to relax and be as positive as you can."



Giant cell tumor of bone

Giant cell tumors are often found at the end of the thigh bone (femur).

Key points

- Giant cell tumor of bone (GCTB) is usually benign (not cancer). The tumor consists of many very large cells or giant cells that can destroy bone.
- When possible, treatment for a giant cell tumor almost always involves surgery to remove the tumor. Treatment aims to prevent damage to the bone near the affected joint.
- Before starting treatment, you'll have tests to see if the tumor is in one area or has spread to distant sites (metastasized). Treatment will be based on whether surgery is possible.
- In metastatic disease, GCTB may spread from the initial tumor to the rest of the body.
- After completing treatment, you'll be monitored to watch for signs that the tumor has returned, called recurrence.
- GTCB often returns to the same area. Treatment is based on whether this is a local or metastatic recurrence.

Questions to ask

- Which treatment(s) do you recommend and why?
- Does this treatment offer a cure? If not, how well can treatment stop the tumor from growing?
- Does the order of treatments matter?
- What side effects can I expect from this treatment?

9
Other resources

53 What else to know

53 What else to do

53 Where to get help

54 Questions to ask

Want to learn more? Here's how you can get additional help.

What else to know

This book can help you improve your cancer care. But it's not the only resource that you have.

You're welcome to receive as much information and help as you need. Many people are interested in learning more about:

- The details of treatment, especially the different types of surgeries and the pros and cons of each type
- Getting financial help
- Finding an oncologist and a surgeon who are experts in bone cancer
- Coping with side effects

What else to do

Your health care center can help you with next steps. It often has on-site resources to help meet your needs and find answers to your questions. Health care centers can also inform you of resources in your community.

In addition to help from your providers, the resources listed in the next section provide support for many people like yourself. Look through the list below and visit the provided websites to learn more about these organizations.

Where to get help

Bone Marrow & Cancer Foundation
bonemarrow.org

Cancer*Care*
cancercare.org

Imerman Angels
mermanangels.org

MedlinePlus
medlineplus.gov

MIB Agents
mibagents.org

National Cancer Institute (NCI)
cancer.gov/types

Northwest Sarcoma Foundation
nwsarcoma.org

Stupid Cancer
Stupidcancer.org

The Alan B. Slifka Foundation
slifkafoundation.org

The Paula Takacs Foundation for Sarcoma Research
paulatakacsfoundation.org

Triage Cancer
triagecancer.org

Questions to ask

- Who can I talk to about help with housing, food, and other basic needs?
- What help is available for transportation, childcare, and home care?
- What other services are available to me and my caregivers?
- How can I connect with others and build a support system?
- Who can I talk to if I don't feel safe at home, at work, or in my neighborhood?



Let us know what you think!

Please take a moment to complete an online survey about the NCCN Guidelines for Patients.
NCCN.org/patients/response



Words to know

ablation
A procedure that uses extreme cold or extreme heat to destroy cancer cells.

biomarker testing
A laboratory test performed on a sample of tissue, blood, or other body fluid to check for certain genes, proteins, or other molecules associated with cancer. Also called molecular testing.

biopsy
A procedure that removes fluid or tissue samples to be tested for a disease.

bone marrow
The soft, sponge-like tissue in the center of most bones where blood cells are made.

bone marrow aspirate
The removal of a small amount of liquid bone marrow to test for disease.

bone marrow biopsy
The removal of a small amount of solid bone and bone marrow to test for disease.

chemotherapy
Drugs that kill fast-growing cells, including cancer cells and normal cells.

chondrosarcoma
Cancer that starts in the cells that form cartilage.

chromosomes
Long strands that contain bundles of coded instructions in cells for making and controlling cells.

clinical trial
A study of how safe and helpful tests and treatments are for people.

complete blood count
A lab test that includes the numbers of different types of blood cells.

CT
A test that uses x-rays from many angles to make a picture of the insides of the body.

contrast
A substance put into your body to make clearer pictures during imaging tests.

core needle biopsy
A procedure that removes tissue samples with a hollow needle. Also called core biopsy.

embolization
A procedure that treats tumors by cutting off their blood supply.

Ewing sarcoma
Cancer that usually occurs in the bones but can emerge in soft tissues (organs).

hypercalcemia
Higher than normal levels of calcium in the blood.

gene
Coded instructions in cells for making new cells and controlling how cells behave.

grade
A rating of how normal or abnormal cancer cells look when viewed under a microscope.

immunotherapy
A treatment with drugs that help the body find and destroy cancer cells.

MRI
A test that uses radio waves and powerful magnets to make pictures of the insides of the body.

metastasis
The spread of cancer from the first tumor to a new site.

monitoring
Testing after treatment ends to check for the return of cancer. Also called surveillance.

oncologist
A doctor who is an expert in the treatment of cancer using medications.

osteosarcoma
Cancer that starts in bone-forming cells called osteoblasts.

palliative care
Health care that includes symptom relief but not cancer treatment.

PET scan
A test that uses radioactive material to detect cancer, heart disease, and brain conditions.

primary treatment
The main treatment used to rid the body of cancer.

primary tumor
The first mass of cancerous cells.

prognosis
The predicted pattern and outcome of a disease.

progression
The growth or spread of cancer after being tested or treated.

radiation therapy
A treatment that uses high-energy rays.

recurrence
The return of cancer after a cancer-free period.

refractory
A cancer that does not improve with treatment.

relapse
The return or worsening of cancer after a period of improvement.

side effect
An unhealthy or unpleasant physical or emotional response to treatment.

soft tissue sarcoma
A cancer that starts in the cells of fat, muscles, nerves, tendons, blood and lymph vessels, and other supportive tissues of the body.

supportive care
Treatment for the symptoms or health conditions caused by cancer or cancer treatment. Also sometimes called best supportive care.

surveillance
Testing after treatment ends to check for the return of cancer. Also called monitoring.

systemic drug therapy
Treatment that works throughout the body.

targeted therapy
A drug treatment that targets and attacks specific cancer cells.

translocation
When pieces of 2 chromosomes (long strands of coded instructions for controlling cells) break off and switch places with each other.

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This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Bone Cancer, Version 1.2026. It was adapted, reviewed, and published with help from the following people:

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Memorial Sloan Kettering Cancer Center
New York, New York

Moffitt Cancer Center
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of Northwestern University
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St. Jude Children's Research Hospital/
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UC Davis Comprehensive Cancer Center
Sacramento, California

UC San Diego Moores Cancer Center
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UCLA Jonsson Comprehensive Cancer Center
Los Angeles, California

UCSF Helen Diller Family Comprehensive Cancer Center
San Francisco, California

University of Colorado Cancer Center
Aurora, Colorado

University of Michigan Rogel Cancer Center
Ann Arbor, Michigan

University of Wisconsin Carbone Cancer Center
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Index

ablation 16, 21, 30–31, 41, 49

amputation 18, 29, 34, 40

appendicular skeleton 39

axial skeleton 39

best supportive care 35

biopsy 9, 12–14, 16, 41, 48

chemotherapy 19–21, 25, 27, 29, 34, 37

clinical trials 22–23

embolization 16, 21, 49

family history 9, 13

fertility 18–19

immunotherapy 19, 25

limb-sparing surgery 18, 29, 40

local control therapy 29

performance status 10

pregnancy 10, 19

radiation therapy 6, 16, 21, 30, 34–37, 40–42, 45–46, 50

radiofrequency ablation 21

radiopharmaceuticals 31

rehabilitation 16

rotationplasty 18

supportive care 21, 23, 31, 35, 41, 45

surgical margin 17

systemic drug therapy 16, 20, 36–37, 40

targeted therapy 19, 20, 25



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