NCCN GUIDELINES FOR PATIENTS®

2025

Bone Cancer

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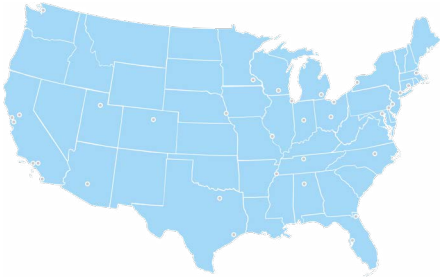
About the NCCN Guidelines for Patients®

National Comprehensive Cancer Network®



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®).

Cancer care is always changing. NCCN develops evidence-based cancer care recommendations used by health care providers worldwide. These frequently updated recommendations are the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®). The NCCN Guidelines for Patients plainly explain these expert recommendations for people with cancer and caregivers.

These NCCN Guidelines for Patients are based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Bone Cancer, Version 1.2025 – August 20, 2024.

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NCCN Foundation seeks to support the millions of patients and their families affected by a cancer diagnosis by funding and distributing NCCN Guidelines for Patients. NCCN Foundation is also committed to advancing cancer treatment by funding the nation's promising doctors at the center of innovation in cancer research. For more details and the full library of patient and caregiver resources, visit NCCN.org/patients.

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1
About bone cancer

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 cancers that can spread (metastasize) to the bone. This chapter presents an overview of bone cancer.

What is bone cancer?

Cancers that start in the bone are called primary bone cancers. Bone cancers that start in another part of the body (such as the breast, lungs, or kidney) and travel to the bones are called secondary bone malignancies or metastatic cancer. Metastatic cancers to bone behave very differently from primary bone cancers.

Sarcomas

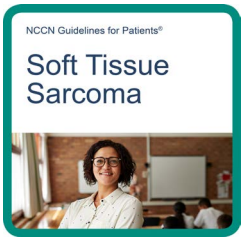
Most primary bone cancers are sarcomas. Sarcomas are a rare group of cancers.

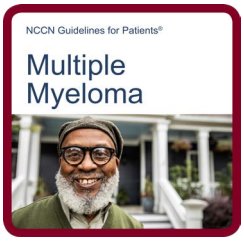
There are 2 main types of sarcomas:

- 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. Osteosarcoma, Ewing sarcoma, and chondrosarcoma are the most common types of bone cancer, except multiple myeloma.

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





Types of bone tumors

A bone tumor is an abnormal growth of cells within a bone. A bone tumor may be cancerous (malignant) or noncancerous (benign). Bone tumors often occur in areas of the bone that grow rapidly (the metaphysis).

This book will discuss the following:

- **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.
- **Chordoma** – starts in notochordal cells that form the spine and vertebrae. Chordomas are usually found in the lower spinal column (sacrum) or at the base of the skull.
- **Ewing sarcoma** – starts in the bone marrow and commonly occurs in the

diaphysis of long bones or flat bones like the pelvis or scapula (shoulder blade).

- **Giant cell tumor of bone (GCTB)** – is an overgrowth of cells found in the part of the bone called the metaphysis and extends into the area of the bone that widens to form a joint (epiphysis).
- **Osteosarcoma** – starts in cells most commonly in the areas of the bone that grow rapidly (metaphysis), near the growth plates.

What is bone?

Bone is a living tissue made up of specialized bone cells and collagen. Collagen is the most abundant protein in the body. Bone tissue also contains blood vessels, nerves, proteins, vitamins, and minerals.

Bone is made up of the following:

- **Compact bone or cortical bone** – the hard, dense outer layer of bone that is covered by a membrane of strong connective tissue called the periosteum
- **Spongey bone or cancellous bone** – the spongy, porous inner layer of bone that contains red marrow
- **Bone marrow** – found in the center of most bones and has many blood vessels

There are 2 types of bone marrow: red and yellow. Red marrow contains blood stem cells that can become red blood cells, white blood cells, or platelets. Yellow marrow is made mostly of fat. Red marrow is often replaced by yellow marrow as we age.

Why you should read this book

Making decisions about cancer care can be stressful. You may need to make tough decisions under pressure about complex choices.

The *NCCN Guidelines for Patients* are trusted by patients and providers. They clearly explain current care recommendations made by respected experts in the field. Recommendations are based on the latest research and practices at leading cancer centers.

Cancer care is not the same for everyone. By following expert recommendations for your situation, you are more likely to improve your care and have better outcomes as a result. Use this book as your guide to find the information you need to make important decisions.

Our body has 206 bones. Bones provide structure and support, protect our organs, create blood cells, store chemicals the body needs, and produce hormones. Bone is light, yet strong and can regrow.

What's in this book?

This book is organized into the following chapters:

Chapter 2: Testing for bone cancer provides an overview of tests you might receive, how fertility might be impacted by treatment, and the role of biomarker and tumor mutation testing.

Chapter 3: Bone cancer staging explains how bone cancer is staged and where bone tumors might be found in the body.

Chapter 4: Types of treatment gives an overview of the types of different treatment and who might be on your treatment team.

Chapter 5: Chondrosarcoma discusses chondrosarcoma treatment. Treatment is usually surgery to remove the tumor.

Chapter 6: Chordoma discusses chordoma treatment. Treatment options include surgery or radiation therapy.

Chapter 7: Ewing sarcoma discusses Ewing sarcoma treatment options. Treatment begins with systemic therapy.

Chapter 8: Giant cell tumor of bone discusses GCTB treatment options. GCTB is usually not cancer, but needs aggressive treatment to prevent bone damage.

Chapter 9: Osteosarcoma discusses osteosarcoma treatment options. Osteosarcoma is the more common type of bone cancer. Treatment is a combination of therapies.

Chapter 10: Other resources provides information on patient advocacy groups and where to get help.

What can you do to get the best care?

Advocate for yourself. You have an important role to play in your care. In fact, you're more likely to get the care you want by asking questions and making shared decisions with your care team. Consider seeking the opinion of a specialist in your type of bone cancer.

The *NCCN Guidelines for Patients* will help you understand cancer care. With better understanding, you'll be more prepared to discuss your care with your team and share your concerns. Many people feel more satisfied when they play an active role in their care.

You may not know what to ask your care team. That's common. Each chapter in this book ends with an important section called *Questions to ask.* These suggested questions will help you get more information on all aspects of your care.

Take the next step and keep reading to learn what is the best care for you!

2
Testing for bone cancer

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 cancer is very rare. Many health care providers are not experts in diagnosing or treating these tumors. Therefore, it is important to find a doctor, treatment center, or hospital that has experience in your type of bone cancer. Use resources on page 61 of this book to help you find an experienced doctor and treatment center.

General health tests

Some general health tests are described next.

Medical history

A medical history is a record of all health issues and treatments you have had in your life. Be prepared to list any illness or injury and when it happened. Bring a list of old and new medicines and any over-the-counter (OTC) medicines, herbals, or supplements you take. Some supplements interact with and affect medicines that your care team may prescribe. Tell your care team about any symptoms you have. A medical history, sometimes called a health history, will help determine which treatment is best for you.

Family history

Your care team will ask about the health history of family members who are blood relatives. This information 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. It's important to know the specific type of cancer or where the cancer started, if it is in multiple locations, and if they had genetic testing.

Physical exam

During a physical exam, your health care provider may:

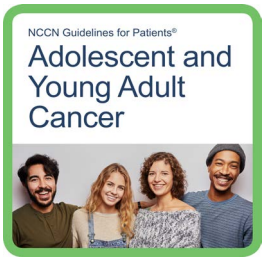
- Check your temperature, blood pressure, pulse, and breathing rate
- Check your height and weight
- Listen to your lungs and heart
- Look in your eyes, ears, nose, and throat
- Feel and apply pressure to parts of your body to see if organs are of normal size, are soft or hard, or cause pain when touched.
- Feel for enlarged lymph nodes in your neck, underarm, and groin
- Examine your skeletal system
- Assess your ability to manage tasks and activities that are used in daily life called a functional assessment

Fertility (all genders)

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.

Fertility preservation is all about keeping your options open, whether you know you want to have children later in life or aren't sure at the moment. Fertility and reproductive specialists can help you sort through what may be best for your situation.

For more information on fertility preservation, see the *NCCN Guidelines for Patients®: Adolescent and Young Adult Cancer* 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. This loss of fertility is related to your age at 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. Therefore, becoming pregnant or having one's partner become pregnant during treatment should be avoided. Hormonal birth control may or may not be recommended, so ask your doctor about options such as intrauterine devices (IUDs) and barrier methods. Types of barrier methods include condoms, diaphragms, cervical caps, and the contraceptive sponge.

Performance status

Performance status (PS) is a person's general level of fitness and ability to perform daily tasks. Your state of general health will be rated using a PS scale called the Eastern Cooperative Oncology Group (ECOG) score or the Karnofsky Performance Status (KPS). PS is one factor taken into consideration when choosing a treatment plan.

Blood tests

Blood tests check for signs of disease 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. Some blood tests are described next.

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 (CBC) measures the levels of red blood cells (RBCs), white blood cells (WBCs), and platelets (PLTs) in your blood. RBCs carry oxygen throughout your body, WBCs fight infection, and PLTs control bleeding.

Comprehensive metabolic panel

A comprehensive metabolic panel (CMP) measures substances in your blood. It provides important information about how well your kidneys and liver are working, among other things. A CMP will look for higher than normal calcium levels in your blood called hypercalcemia. Calcium is needed for healthy teeth, bones, and other body tissues. Bone damage from cancer can cause your bones to release calcium into the bloodstream, which can lead to illness and other health issues.

Lactate dehydrogenase

Lactate dehydrogenase (LDH) or lactic acid dehydrogenase is an enzyme found in most cells. Dying cells release LDH into blood. Fast-growing cells, such as tumor cells, also release LDH.

Pregnancy test

Those who can become pregnant should be given a pregnancy test before treatment begins.

Imaging tests

Imaging tests take pictures of the inside of your body to look for cancer deposits. A medical doctor called a radiologist will interpret the test and send a report to your doctor. While these reports might be available to you through your patient portal or patient access system, please wait to discuss these results with your care team. You may not have all of the following tests.

X-ray

An x-ray is a type of radiation. In small doses, it is used to make pictures of the inside of the body. It might be referred to as a radiograph.

Bone scan

A bone scan uses a radiotracer. A radiotracer is 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.

A special camera will take pictures of the tracer in your bones as it moves over your body. Areas of bone damage take up more radiotracer than healthy bone and 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 will be asked to hold still during the scan. At one point the scanner will be less than one inch from your face. Talk to your care team about what to expect.

Contrast material

Contrast material is used to improve the quality of the pictures of the inside of the body. Contrast materials are substances that help enhance and improve the images of several organs and structures in the body. It is used to make the pictures clearer. The contrast is not permanent and will leave your body in your urine immediately after the test. The types of contrast vary and are different for CT and MRI.

Tell your care team if you have had allergic reactions to contrast in the past. This is important. You might be given medicines to avoid the effects of those allergies. Contrast might not be used if you have a serious allergy or if your kidneys aren't working well.

CT scan

A CT or CAT (computed tomography) scan uses x-rays and computer technology to take pictures of the inside of the body. It takes many x-rays of the same body part from different angles. All the images are combined to make one detailed picture. In many cases, contrast will be used.

MRI scan

An MRI (magnetic resonance imaging) scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays, which means there is no radiation delivered to your body during the test. Because of the very strong magnets used in the MRI machine, tell the technologist if you have any metal in your body. During the test, you will likely be asked to hold your breath for 10 to 20 seconds as the technician collects the images. Contrast is often used.

A closed MRI has a capsule-like design where the magnet surrounds you. An open MRI has a magnetic top and bottom, which allows for an opening on each end. Closed MRIs are more common than open MRIs, so if you have claustrophobia (a dread or fear of enclosed spaces), be sure to talk to your care team about it.

PET scan

A PET (positron emission tomography) scan uses a radioactive drug called a tracer. A tracer is a substance injected into a vein to see where cancer cells are in the body and how much sugar is being taken up by the cancer cells. This gives an idea about how fast the cancer cells are growing. Cancer cells show up as bright spots on PET scans. However, not all tumors will appear on a PET scan. Also, not all bright spots found on the PET scan are cancer. It is normal for the brain, heart, kidneys, and bladder to be bright on PET. Inflammation or infection can also show up as a bright spot. When a PET scan is combined with CT, it is called a PET/CT scan.

An FDG-PET/CT uses a radiotracer called fluorodeoxyglucose (FDG). It is made of

fluoride and a simple form of sugar called glucose. You cannot eat or drink for at least 4 hours before the scan. This scan is most helpful when other imaging results are unclear.

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 core needle or surgical biopsy. The placement of the biopsy is critical. Therefore, it is recommended that the biopsy be performed by an experienced surgeon at the center where you will receive treatment.

- **Core needle biopsy** removes tissue samples with a hollow needle that can grab a piece of tissue. This is frequently performed under light sedation by an interventional radiologist. It can also be performed by your surgeon in the office.
- **Surgical (open) biopsy** removes a sample of the tumor through a small incision. A surgical biopsy should be performed by your treating surgeon. This requires a trip to the operating room.

Bone marrow tests

Bone marrow tests might be done in certain cases.

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

- Bone marrow aspirate
- Bone marrow biopsy

It is recommended that a biopsy be performed by an experienced surgeon at the cancer center where you will receive treatment.

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 will likely lie on your belly or side. Your doctors will first clean and give sedation and/or numb your skin and outer surface of your bone. 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 the biopsy, a wider needle will be used to remove a core sample. You may feel bone pain at your hip for a few days. Your skin may bruise.

Genetic cancer risk testing

Genetic testing is done using blood or saliva from spitting into a cup or a cheek swab. The goal is to look for gene mutations inherited from your birth parents called germline mutations. Some mutations can put you at risk for more than one type of cancer. You can pass these genes on to your children. Also, blood relatives might carry these mutations. Tell your care team if there is a family history of cancer.

Biomarker testing

A sample from your biopsy will undergo lab tests to look for specific DNA mutations/alterations, protein levels, or other molecular features. 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 that are important for you.

Biomarker testing includes tests of genes or their products (proteins). It identifies the presence or absence of mutations and certain proteins that might suggest treatment. Proteins are written like this: IDH1. Genes are written with italics like this: *IDH1*. When a gene or protein is found, it is shown with a plus sign (+) like this: IDH1+. When a gene or protein has not been found, it is written with a negative sign (-) like this: IDH1-.

Ewing sarcoma

A cell must make a copy of its chromosomes before dividing into two cells. Sometimes, there are mistakes in the copies. One type of mistake is when parts (DNA) of two chromosomes break off and switch with each other. This is called a translocation. It can result in a fusion of two genes. A translocation between chromosomes 11 and 22 is written as t(11;22) and is common in Ewing sarcoma.

Ewing sarcoma is characterized by the fusion of the *EWS* gene (*EWSR1*) on chromosome 22 with various members of the *ETS* gene family (*FLI1, ERG, ETV1, ETV4,* and *FEV*), 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.

MSI-H/dMMR mutation

Microsatellites are short, repeated strings of DNA. When errors or defects occur, they are fixed by mismatch repair (MMR) proteins. Some cancers have DNA mutations or changes that prevent these errors from being fixed. This is called microsatellite instability (MSI) or deficient mismatch repair (dMMR). When cancer cells have more than a normal number of microsatellites, it is called MSI-H (microsatellite instability-high). This is often due to dMMR genes. Pembrolizumab (Keytruda) is often used to treat MSI-H/dMMR tumors.

Tumor mutational burden

When there are 10 or more mutations per million base pairs of tumor DNA, it is called tumor mutational burden-high (TMB-H). Pembrolizumab (Keytruda), nivolumab (Opdivo), or ipilimumab (Yervoy) might be used for TMB-H tumors in some cases.

Tumor mutation testing

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

Mutation testing uses methods such as next-generation sequencing (NGS) and polymerase chain reaction (PCR).

IDH1 mutation

Some gene mutations such as *IDH1* can be targeted with specific therapies such as ivosidenib (Tibsovo). Ivosidenib is for those with chondrosarcoma who are prone to have *IDH1* mutations.

Next-generation sequencing

Next-generation sequencing (NGS) is a method used to determine a portion of a person's DNA sequence. It shows if a gene has any mutations that might affect how the gene works. NGS looks at the gene in a more detailed way than other methods and can find mutations that other methods might miss.

PCR

A polymerase chain reaction (PCR) is a technique that can make millions or billions of copies of your DNA or RNA (genetic information). PCR is very sensitive. It can find 1 abnormal cell among more than 100,000 normal cells. These copies, called PCR products, might be used for NGS.

"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."

Key points

- Tests are used to plan treatment and check how well treatment is working.
- Some types of treatment for bone cancer can affect fertility. Those who want to have children in the future should be referred to a fertility specialist before starting treatment.
- A diagnosis of bone cancer is confirmed using a core needle or surgical biopsy. The placement of the biopsy is critical. Therefore, it is recommended that the biopsy be performed at an experienced surgeon at the center where you will receive treatment.
- A sample from a biopsy of your tumor might be tested to look for biomarkers or proteins.
- 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?
- 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?

3
Bone cancer staging

Cancer staging is used to help make treatment decisions. It describes the size and location of the tumor, and if cancer has spread to lymph nodes, organs, or other parts of the body. This chapter provides a general overview of cancer staging.

Overview

There are many staging systems. Most staging systems include information about:

- Where the tumor is located in the body
- The size of the tumor
- Whether the cancer has spread to nearby lymph nodes
- Whether the cancer has spread to a different part of the body
- Tumor grade, which refers to how abnormal the cancer cells look under a microscope

How is bone formed?

Bone is a living tissue made of cells (osteocytes, osteoblasts, and osteoclasts), calcium, phosphate, hydroxyapatite (a bone mineral), and collagen. New bone formation, known as ossification, starts in the womb and ends during adolescence, between 10 and 19 years of age. Ossification is complete when the skeleton is done growing.

Once ossification is complete, bone is continuously being replaced by a process called remodeling. Remodeling breaks down and rebuilds bone and helps maintain normal calcium levels in the body. When calcium is needed, cells called osteoclasts break down bone. This releases calcium into the bloodstream. At the same time, bone-forming cells called osteoblasts create bone from calcium in the bloodstream. Some types of bone cancer can interrupt this process.

Parts of bone:

- **Epiphysis** – the end of the bone, which is covered with cartilage to make a joint.
- **Metaphysis** – located between the physis (growth plate) and diaphysis.
- **Diaphysis** – the middle region of the bone.
- **Physis** – the growth plate, which is made of cartilage. After skeletal maturity, this leaves a physeal scar or line visible in the bone.

TNM scores

The tumor, node, metastasis (TNM) system is used to stage most bone cancers. In this system, the letters T, N, and M describe different areas of cancer growth. Based on imaging and biopsy results, your doctor will assign a score or number to each letter. The higher the number, the larger the tumor or the more the cancer has spread to lymph nodes or other organs. These scores will be combined to assign the cancer a stage. A TNM example might look like this: T1N0M0 or T1, N0, M0. This might represent a stage 1 cancer.

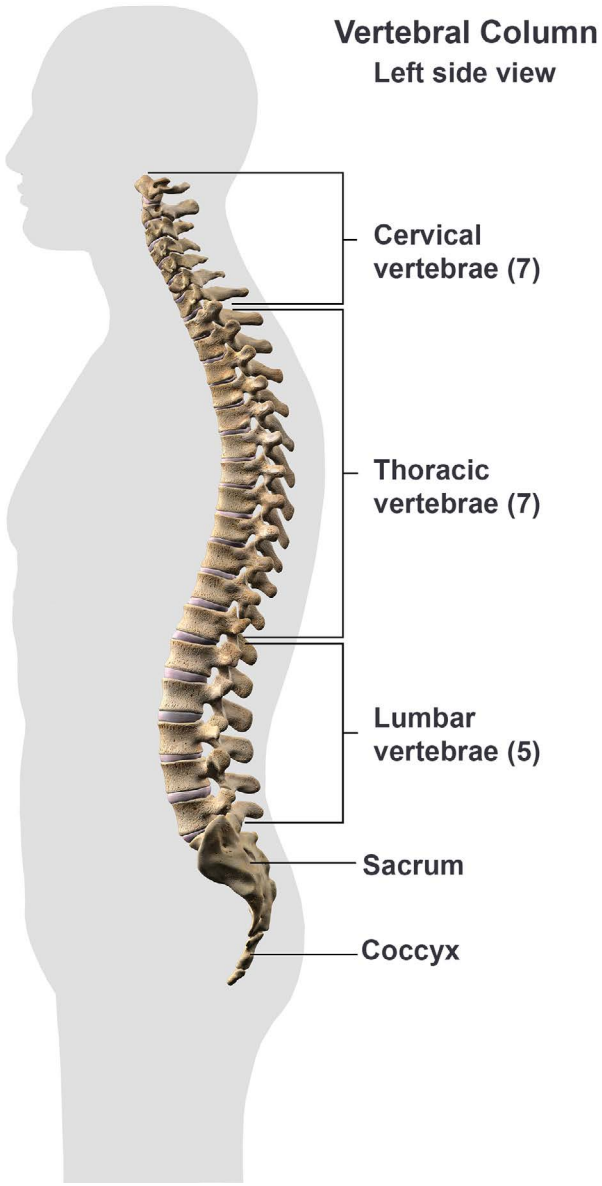
- **T (tumor)** – Size and depth of the main (primary) tumor
- **N (node)** – If cancer has spread to nearby (regional) lymph nodes
- **M (metastasis)** – If cancer has spread (metastasized) to distant parts of the body

Cancer staging is often done twice.

- **Clinical stage (c)** is the rating given before any treatment. It is based on a physical exam, biopsy, and other tests.
- **Pathologic stage (p)** or surgical stage is determined by examining tissue removed during surgery such as in the removal of a lymph node. Pathologic features include size, shape, and type of cell.

The spine

The spinal or vertebral column includes a flexible column of bones called vertebrae. The spine includes the sacrum and tailbone (coccyx). There are 24 bones in the flexible spine: 7 cervical (neck), 12 thoracic (upper back), and 5 lumbar (lower back).



T = Tumor

The primary (main) tumor size is measured in centimeters (cm). A large pea is 1 cm. A golf ball is 4 cm. A baseball is 7 cm. A grapefruit is 15 cm.

Tumors in limbs, torso, skull, and face

Tumors are staged based on location. The appendicular skeleton is composed of 126 bones of the upper limbs, the lower limbs, the shoulder girdle, and the pelvic girdle. The shoulder girdle is a set of bones that connects the arm to the body. The pelvic girdle is a set of bones that connects the leg to the body.

Staging for tumors found in the appendicular skeleton, torso, skull, and face are described next.

- **TX** – Primary tumor cannot be measured
- **T0** – Tumor cannot be found
- **T1** – Tumor is 8 cm or smaller
- **T2** – Tumor is larger than 8 cm
- **T3** – More than 1 tumor in the primary bone site

Tumors in spine

Staging for tumors found in the bones of the spine (vertebra) are described next.

- **TX** – Primary tumor cannot be measured
- **T0** – Tumor cannot be found
- **T1** – Tumor confined to 1 vertebra or 2 vertebrae next to one another (adjacent)
- **T2** – Tumor confined to 3 adjacent vertebrae
- **T3** – Tumor confined to 4 or more adjacent vertebrae, or any nonadjacent vertebrae
- **T4** – Tumor has grown into spinal canal or large (great) vessels

Tumors in pelvis

Staging for tumors found in the pelvis are described below.

- **TX** – Primary tumor cannot be measured
- **T0** – Tumor cannot be found
- **T1** – Tumor is confined to 1 pelvic segment and has not grown outside the bone
  - **T1a** – Tumor is 8 cm or less
  - **T1b** – Tumor is larger than 8 cm
- **T2** – Tumor is confined to 1 pelvic segment but has grown outside the bone or 2 segments that have not grown outside the bone
  - **T2a** – Tumor is 8 cm or less
  - **T2b** – Tumor is larger than 8 cm
- **T3** – Tumor spans 2 pelvic segments and has grown outside the bone
  - **T3a** – Tumor is 8 cm or less
  - **T3b** – Tumor is larger than 8 cm
- **T4** – Tumor spans 3 pelvic segments or crosses the sacroiliac joint (SIJ)
  - **T4a** – Tumor involves SIJ and extends into the spine

- **T4b** – Tumor has grown into major blood vessels

N = Node

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are those located near the tumor. Cancer in the lymph nodes is called nodal disease and is rare in bone cancer.

- **NX** – Cancer in nearby lymph nodes cannot be measured.
- **N0** – There is no cancer in nearby lymph nodes.
- **N1** – Cancer is found in the regional lymph nodes.

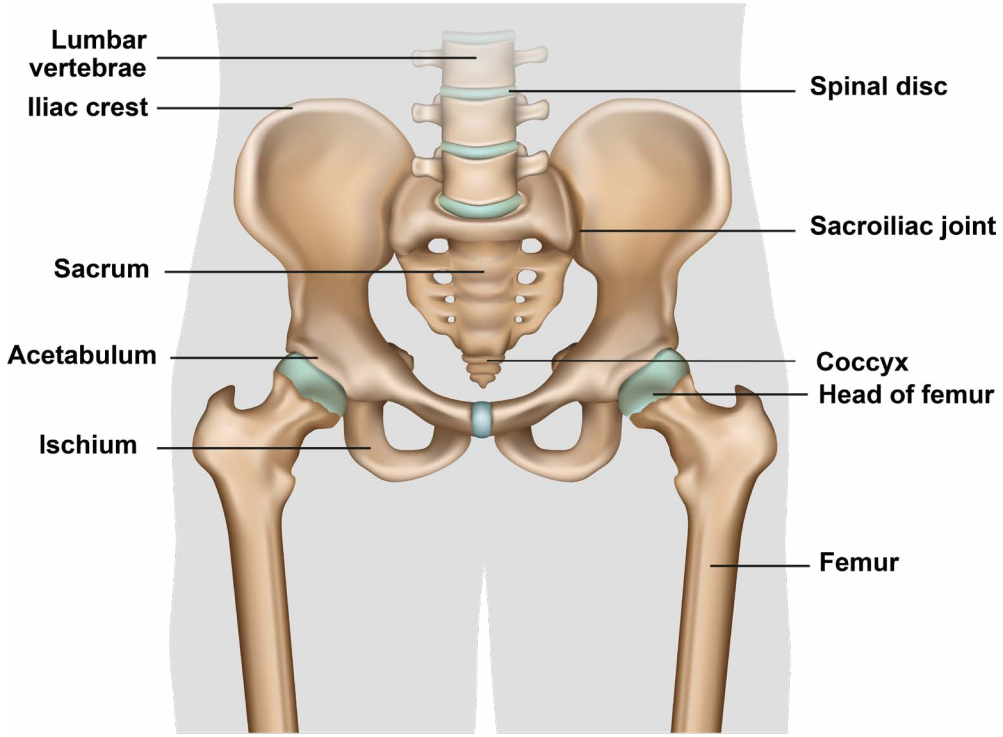
M = Metastasis

Bone cancer can spread through blood or the lymphatic system. Cancer that has spread to distant parts of the body is defined as M1. When cancer is not metastatic it is labeled as M0. Bone cancer can spread to the lung, other bones, or other parts of the body. Bone cancer that has metastasized to the lung is still called bone cancer.

- **M0** – No distant metastasis
- **M1** – Distant metastasis is found
  - **M1a** – Metastasis is in the lung
  - **M1b** – Metastasis is in the bone or other distant sites

The pelvis

The pelvis is a basin-shaped structure that supports the spinal column and protects the abdominal organs.



G = Grade

Another factor used in staging is the cancer grade. Grade describes how abnormal the tumor cells look under a microscope (histology). Higher-grade cancers tend to grow and spread faster than lower-grade cancers. GX means the grade can't be determined, followed by G1, G2, and G3. Well differentiated (G1) means the cancer cells look like normal cells. Poorly differentiated (G3) means the cancer cells look very different compared to normal cells.

- **GX** – Grade cannot be determined
- **G1** – Well differentiated (low grade)
- **G2** – Moderately differentiated (high grade)
- **G3** – Poorly differentiated (high grade)

Numbered stages

Number stages range from stage 1 to stage 4, with 4 being the most advanced. The stage of a bone cancer is determined by combining the T, N, M, and G scores. These stages are written as stage I, stage II, stage III, and stage IV. Not all bone cancers are described using numbered stages. **See Guide 1.**

Guide 1
Bone cancer stages

Stage 1A	• T1, N0, M0, G1 or GX
Stage 1B	• T2 or T3, N0, M0, G1 or GX • T3, N0, M0, G1 or GX
Stage 2A	• T1, N0, M0, G2 or G3
Stage 2B	• T2, N0, M0, G2 or G3
Stage 3	• T3, N0, M0, G2 or G3
Stage 4A	• Any T, N0, M1a, Any G
Stage 4B	• Any T, N1, Any M, Any G • Any T, Any N, M1b, Any G

Key points

- Staging is often used to make treatment decisions. Staging describes how much cancer is in your body, where it is located, and what subtype you have.
- The tumor, node, metastasis (TNM) system is commonly used to stage cancer.
- Regional lymph nodes are found near the primary tumor.
- Cancer that has spread to distant parts of the body is called metastatic cancer.
- The most common site for metastasis of bone cancer is the lung, followed by other bones.

Questions to ask

- Where is the tumor located and what size is the tumor?
- Is there more than one known cancer site?
- Is there cancer in the lymph nodes?
- What stage and grade is the cancer?
- What does the cancer stage and grade mean in terms of treatment and prognosis?

4
Types of treatment

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 (MDT). An MDT is a team of health care and psychosocial care professionals from different professional backgrounds who have knowledge (expertise) and experience in your type of cancer. This team is united in the 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 only 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 any area of the body except the neck and skull and inside the chest and abdomen.
- **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 type of care, the team might include the following specialists:

- **A thoracic surgeon** performs operations within the chest.
- **A plastic surgeon** performs operations to improve function or reconstruct missing areas.
- **An interventional radiologist** performs needle biopsies, ablation therapies, and embolizations, and places ports for systemic therapy.
- **A physiatrist** is a medical doctor 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 patient.
- **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. They may be an orthopedic oncologist.

Other team members might include a physical or occupational therapist, prosthetist, and mental health expert.

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 primary (main) site. Sometimes, surgery can be used to remove limited areas of metastatic disease.
- Relieve pain or discomfort

The goal of surgery is to remove all of the cancer. To fully remove all of the cancer, your surgeon must cut out the tumor with a rim of normal tissue around the edge. This allows the pathologist to determine a surgical margin. A clear or negative margin resection (R0) is when no cancerous cells are found in the tissue around the edge of the tumor. An R1 positive margin or R1 resection indicates the surgeon has removed all of the visible tumor, but the microscopic margins are still positive for tumor cells. In an R2 positive margin, the surgeon does not or is unable to remove all of the visible tumor.

A tumor that can be removed completely with surgery is called resectable. Not all tumors are resectable. You might receive treatment to shrink a tumor or the amount of cancer before surgery. This is called preoperative or neoadjuvant treatment. Treatment after surgery might be referred to as postoperative or adjuvant treatment.

"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."

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

When preparing for surgery, seek care or ask for a referral to a hospital or cancer center that has experience in treating your type of bone cancer. It is important to find an experienced surgeon who has performed this type of procedure many times with good results.

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 (LSS) or limb salvage surgery.

Amputation

Amputation is surgery to remove part or all of a limb such as an arm or leg. There are different types of amputations. Some types include reconstructive surgery, bone grafts, or implanted pieces.

Rotationplasty

In a rotationplasty, the bottom of the thigh bone (femur), the knee, and the 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 allow knee function when it otherwise would not be possible. Not all surgeons or cancer centers offer this type of surgery.

Rehabilitation after surgery

Rehabilitation, such as physical and occupational therapy, will be part of any limb-sparing surgery, amputation, rotationplasty, or reconstructive surgery. Rehabilitation may include occupational therapy to help with daily life skills or physical therapy to help your body move and function. A physiatrist is a medical doctor who specializes in rehabilitation.

Seek a peer support group to connect with others who have experienced the loss of 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 metastases that are causing pain. Surgery to remove a metastasis is called a metastasectomy. This is different from a mastectomy, which is surgery to remove the breast. Palliative surgery is sometimes offered to fix a broken bone or help prevent a cancerous bone from breaking.

Systemic therapy

Systemic (drug) therapy works throughout the body. Types include chemotherapy, targeted therapy, and immunotherapy. Systemic therapy might be used alone or with other therapies. Goals of systemic therapy should be discussed before starting treatment. Your wishes about treatment are important. Make your wishes known.

- **Neoadjuvant or preoperative therapy** is systemic therapy or radiation given before surgery.
- **Perioperative therapy** is systemic therapy given before and after surgery.
- **Adjuvant or postoperative therapy** is systemic therapy or radiation therapy given after surgery.
- **Palliative therapy** might be the term used for systemic therapy given for advanced or metastatic disease.

Systemic therapies are often described in the following ways:

- **Preferred therapies** have the most evidence they work better than others.
- **Other recommended therapies** may not work quite as well as preferred therapies, but they can still be useful.
- **Therapies used in certain** cases are for people 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. When only one drug is used, it's called a single agent. A combination or multi-agent regimen is the use of two or more chemotherapy drugs.

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. The final dose differs between people because it is based on body weight and height. Intrathecal (IT) chemotherapy is injected into spinal or brain fluid.

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.

Targeted therapy

Targeted therapy is a form of systemic therapy that focuses on specific or unique features of cancer cells. Targeted therapies seek out how cancer cells grow, divide, and move in the body. These drugs stop the action of molecules that help cancer cells grow and/or survive. 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. Others target hormones.

Immunotherapy

Immunotherapy is 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 (RT) uses high-energy radiation from photons, electrons, or protons, and other sources to kill cancer cells and shrink tumors. It is given over a certain period of time. Radiation therapy can be given alone or with certain systemic therapies. When used as supportive care to help ease pain or discomfort caused by cancer, it is called palliative RT.

Radiation may be given:

- **As the primary (main) treatment**
- **Before surgery,** called neoadjuvant RT, to shrink the tumor before surgery
- **After surgery,** called adjuvant RT, to kill any cancerous cells that remain
- **As palliative treatment** to ease pain or discomfort caused by bone tumors

External beam radiation therapy

External beam radiation therapy (EBRT) uses a machine outside of the body to aim radiation at the tumor(s). There is more than one type of EBRT used in the treatment of bone cancer. These allow for safer, higher doses of radiation.

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

- **Stereotactic body radiation therapy (SBRT)** uses high-energy radiation beams to treat cancers in 5 or fewer treatments.
- **Stereotactic radiosurgery (SRS)** uses special equipment to position the body and give one precise, large dose of radiation.
- **Three-dimensional conformal radiation therapy (3D-CRT)** uses computer software and CT images to aim beams that match the shape of the tumor.
- **Intensity-modulated radiation therapy (IMRT)** uses small beams of different strengths to match the shape of the tumor.
- **Particle beam radiation therapy** uses protons, carbon ions, or other heavy ions to treat bone cancer.

Internal radiation

Internal radiation called brachytherapy is placed inside the body as a solid implant similar to beads or seeds.

Radiopharmaceuticals

Radiopharmaceuticals, such as radium-233 (Xofigo), contain a radioactive substance that emits radiation. This radioactive substance is different than contrast material used in imaging.

Other treatments

Ablation and embolization are described next.

Ablation

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

There are 2 types of thermal ablation used to destroy cancer cells:

- Cryotherapy or cryosurgery
- Radiofrequency ablation (RFA)

Both types of ablation use a special needle, called a probe, that is inserted into the tumor. With cryotherapy, a medical gas is passed through the probe to cause below-freezing temperatures. This freezes the tumor to destroy it. With RFA, the probe emits radio waves to heat the tumor and destroy it. The probe can be guided into place with a CT scan, ultrasound, or other imaging tests. The probe will be removed when treatment is done.

Embolization

Embolization treats tumors by cutting off their blood supply. To do this, a catheter is inserted into an artery and guided to the tumor. Once in place, beads are inserted to block the blood flow. With chemoembolization, the beads are coated with chemotherapy. Radioembolization uses small radioactive beads.

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.

Clinical trials

A clinical trial is a type of medical research study. After being developed and tested in a lab, potential new ways of treating 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. Food and Drug Administration (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 doctor 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.

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 is common to receive either a placebo with a standard treatment, or a new drug with a standard treatment. You will be informed, verbally and in writing, if a placebo is part of a clinical trial before you enroll.

Are clinical trials free?

There is 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 will continue to receive standard cancer care. This care is often covered by insurance.

General supportive care

Supportive care helps improve your quality of life during and after cancer treatment. The goal is to prevent or manage side effects and symptoms, like pain and cancer-related fatigue. It also addresses the mental, social, and spiritual concerns faced by those with cancer.

Supportive care is available to everyone with cancer and their families, not just those at the end of life. Palliative care is another name for supportive care.

Supportive care can also help with:

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

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

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

Late effects

Late effects are side effects that occur months or years after a disease is diagnosed or after treatment has ended. Late effects may be caused by cancer or cancer treatment. They may include physical, mental, and social health issues, and 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. It is important to keep any follow-up doctor visits and imaging test appointments. Seek good routine medical care, including regular doctor visits for preventive care and cancer screening.

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

Key points

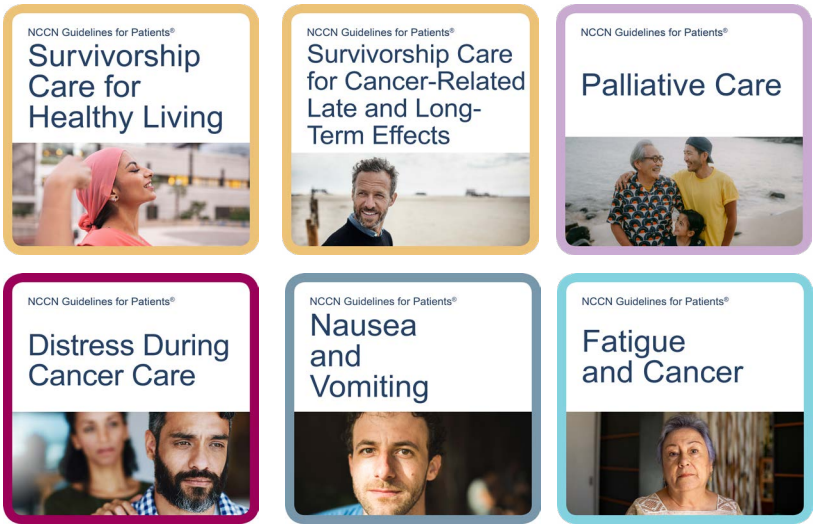
- Surgery is an operation or procedure to remove cancer from the body. The type of surgery depends on the size, location, and number of tumor(s).
- Systemic therapy works throughout the body. It includes chemotherapy, targeted therapy, and immunotherapy.
- Radiation therapy (RT) uses high-energy radiation from 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 doctor, 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?

Supportive care resources

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



5
Chondrosarcoma

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 that is best for you.

Overview

Chondrosarcoma starts in the cells that form cartilage. It is most commonly found in the thigh bone (femur), arm (humerus), or pelvis. Treatment is based on the tumor type, grade, and location.

A tumor that can be removed with surgery is called resectable. An unresectable tumor cannot be removed with surgery. Surgery to remove all of the tumor is called wide excision. Intracompartmental tumors have not grown outside the bone. Extracompartmental tumors have grown outside of the bone.

Treatment is grouped as follows:

- **Locally aggressive tumors**
- **Malignant tumors** which include
  - Low-grade extracompartmental appendicular tumors have grown outside the bone in the appendicular skeleton.
  - Grade 1 axial tumors occur in the axial skeleton, which includes the pelvis, ribs, sternum, and scapula.
  - High-grade (grade 2 or 3) tumors look very abnormal under a microscope.
  - Clear cell tumorsare a type of high-grade chondrosarcoma.
  - Extracompartmental tumors have grown through the bone wall and out of the area where they started.
- **Metastatic disease**
- **Dedifferentiated tumors** are treated with osteosarcoma therapy
- **Mesenchymal tumors** are treated with Ewing sarcoma therapy

Treatment for primary tumors

Treatment options will be based on the type and location of the chondrosarcoma, your wishes, and your care team's recommendations.

Locally aggressive tumors

This section is for treatment of atypical cartilaginous tumors (ACTs), which are low-grade intracompartmental tumors of the appendicular skeleton. The appendicular skeleton includes the shoulder girdle, the upper limbs, the pelvic girdle, and the lower limbs. These tumors have not grown through the bone wall or spread to any other part of the body. ACTs rarely metastasize and that is why they are called locally aggressive tumors. In some cases, ACTs can transform into a malignant, more aggressive tumor. If this happens, it will be treated as a cancer.

Depending on the location of the tumor, treatment is intralesional excision (surgery) to remove part of the tumor. This is also called curettage. Intralesional excision is not an option for pelvic tumors or tumors determined to be malignant. Surgery to remove all of the tumor (wide excision) might be an option.

After completing treatment, you will be monitored with the following tests for the return of the tumor called recurrence.

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

Malignant tumors

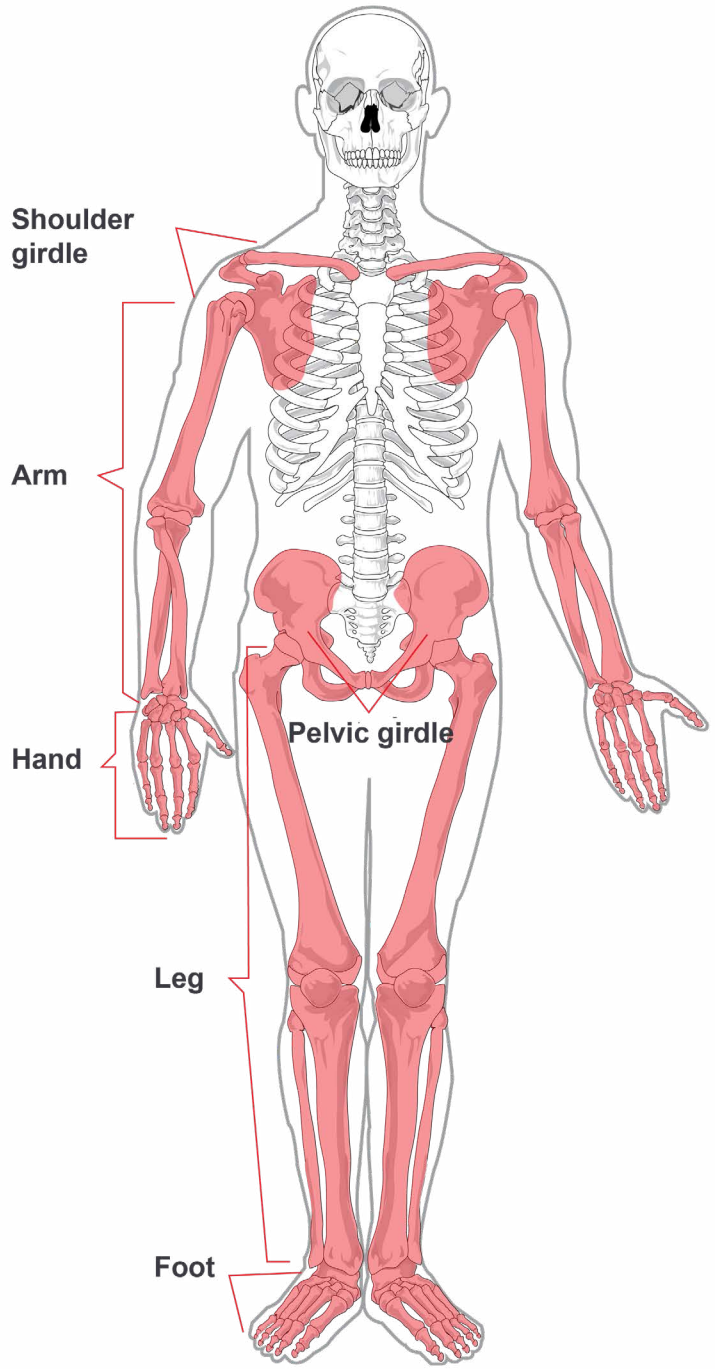
Malignant tumors are cancers that tend to metastasize and spread. Therefore, they need more aggressive treatment than benign, locally aggressive tumors.

Treatment options include:

- If resectable, surgery to remove all of the tumor (wide excision). This might be limb-sparing excision or limb amputation.
- If borderline resectable or unresectable, radiation therapy might be given.

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.



After treatment you will have the following to monitor for recurrence:

- Physical exam
- X-rays of primary site, and CT and MRI scans as needed
- Chest imaging 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
- Re-assess function at every follow-up visit

Recurrence

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

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

- If resectable, surgery to remove all of the tumor (wide excision). If surgery does not remove all of the cancer, then you might have more surgery or radiation therapy.
- If unresectable, radiation therapy might be given.

Metastatic disease

Treatment for metastatic chondrosarcoma is based on the amount of observed metastatic disease. If there are a few sites of metastasis, this is termed oligometastatic. If there are many sites, then this is called widespread metastasis. Treatment focuses on reducing the number of metastases or amount of cancer.

Oligometastases

In oligometastases, there are a limited number of metastases. If possible, you will have surgery to remove all of the metastases. Radiation therapy is an option for unresectable metastases. A clinical trial using systemic therapy is also an option.

Widespread disease

Treatment options for widespread disease might include:

- Radiation therapy, surgery, and/or ablation therapies for sites causing symptoms
- Systemic therapy such as dasatinib (Sprycel) or pazopanib (Votrient)
- Clinical trial

Before starting systemic therapy, biomarker testing might be done. Not everyone with metastatic chondrosarcoma is able to have surgery or systemic therapy. For tumors with *IDH1* mutations, ivosidenib (Tibsovo) might be an option. For MSI-H/dMMR tumors, pembrolizumab (Keytruda) is the preferred systemic therapy option. Preferred therapies have the most evidence they work better than others.

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.
- A tumor that can be removed with surgery is called resectable. A tumor that cannot be removed with surgery is called unresectable. Unresectable tumors are often treated with radiation therapy (RT).
- Surgery to remove part of a tumor is called intralesional excision or curettage, and is limited to tumors that are thought to be non-cancerous. Surgery to remove all of a tumor is called wide excision.
- 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.
- A clinical trial is an option for metastatic chondrosarcoma.

Questions to ask

- Is the tumor resectable or unresectable 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?

6
Chordoma

Chordoma is a slow-growing sarcoma that usually starts in the lower spinal column or at the base of the skull. It often grows into the soft tissue around the bone making it difficult to treat. Treatment options include surgery or radiation therapy. Together, you and your care team will choose a treatment plan that is best for you.

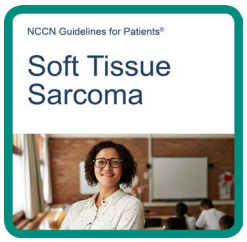
Overview

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.

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, or recur, 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.

This chapter is for conventional or chondroid chordomas. Those with chordoma should be evaluated and treated by a multidisciplinary team who are experts in the management of chordoma. Before starting treatment, you will have imaging tests of the main (primary) sarcoma site.

For those with poorly differentiated or dedifferentiated chordoma, see the *NCCN Guidelines for Patients®: Soft Tissue Sarcoma* at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Treatment

Chordomas are treated based on the location of the tumor:

- Tumors in the spine that include the sacrum and coccyx (sacrococcygeal)
- Tumors at or in the base of the skull

A tumor that can be removed with surgery is called resectable. An unresectable tumor cannot be removed with surgery and might be treated with radiation therapy (RT). Chordomas often grown outside the bone into surrounding tissue near vital organs, veins, and arteries. Parts of these tissues might also need to be removed and sewn back together. This is called resection. The goal is to remove as much of the tumor as possible.

Tumors in spine

For tumors in the spine:

- For a resectable tumor, surgery (wide excision) is the most important part of treatment. RT might be given before, during, or after surgery.
- For an unresectable tumor, RT without surgery might be an option.

Tumors in skull base

For tumors at or in the base of the skull:

- If the tumor is resectable, then surgery (intralesional excision) will be performed. RT might be given before, during, or after surgery. A follow-up MRI with contrast will be done to see if RT or another surgery is needed.
- If the tumor is unresectable, then RT might be an option.

Monitoring

After completing treatment, you will be monitored to watch for signs that cancer has returned called a recurrence. Monitoring includes imaging tests for up to 10 years or as needed.

Recurrence

When cancer returns, it is called a recurrence. Treatment is based on if the recurrence is local or metastatic. 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 might include one or more of the following:

- Surgery
- Systemic 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 in combination. Systemic therapy is drug therapy that works throughout the body. Before starting systemic therapy, biomarker testing might be done. Most systemic therapy options for chordoma recurrence are targeted therapies such as imatinib (Gleevec), dasatinib (Phyrago, Sprycel), and sunitinib (Sutent). Other targeted therapies might be given.

Key points

- Chordomas are treated based on the location of the tumor.
- A tumor that can be removed with surgery is called resectable. The goal of surgery is to remove as much of the tumor as possible.
- An unresectable tumor cannot be removed with surgery and might be treated with radiation therapy (RT).
- When cancer returns, it is 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.
- Best supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life.

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

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 treatment matter?
- What side effects can I expect from this treatment?

7
Ewing sarcoma

Ewing sarcoma is a group of cancers that start in the bone or soft tissue. Treatment begins with systemic therapy. Together, you and your care team will choose a treatment plan that is best for you.

Overview

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

There are 2 types of Ewing sarcoma:

- Ewing sarcoma of bone
- Ewing sarcoma of soft tissues

Ewing sarcoma of bone usually occurs in the thigh (femur), pelvis, and bones of the chest. However, any bone may be affected. When arising in a long bone, the diaphysis is the most common site. Ewing sarcoma can spread to other parts of the body (metastasize), usually to the lungs, other bones, or bone marrow.

Testing

Before starting treatment, you will have imaging, bloodwork, and other tests. Treatment can alter fertility. Therefore, you might be referred to a fertility specialist to discuss fertility preservation options before starting treatment.

For possible tests, see **Guide 2.**

Guide 2
Possible tests for Ewing sarcoma

Medical history and physical exam
MRI with or without CT (both with contrast) of primary site
Chest CT
FDG-PET/CT (preferred) and/or bone scan
Possible bone marrow biopsy and/or screening MRI of spine and pelvis
Biomarker testing (may require re-biopsy)
Lactate dehydrogenase (LDH)
Consider speaking to a fertility specialist before starting treatment

Primary treatment

Systemic therapy is the first or primary treatment for all types of Ewing sarcoma. It will include a combination of chemotherapies. This is called multi-agent chemotherapy. You will have at least 9 weeks of multi-agent first-line chemotherapy.

Systemic therapies are often described in the following ways:

- **Preferred therapies** have the most evidence they work better than others.
- **Other recommended therapies** may not work quite as well as preferred therapies, but they can still be useful.
- **Therapies used in certain** cases are for people with specific cancer features or health circumstances.

For first-line systemic therapy options, **see Guide 3.**

Restaging

After primary treatment of first-line systemic therapy, your cancer will be restaged.

Possible tests to restage 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
- Earlier imaging scans might be repeated

Restaging will determine if the cancer is:

- Stable or improved
- Not responding to treatment or has progressed

Guide 3
First-line systemic therapy options: Ewing sarcoma

Preferred option	• Vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide (VDC/IE). Dactinomycin can be substituted for doxorubicin.
Other recommended	• Vincristine, doxorubicin, ifosfamide, and dactinomycin (VAIA) • Vincristine, ifosfamide, doxorubicin, and etoposide (VIDE)

*Note: Dactinomycin may be substituted for doxorubicin

Stable or improved disease

This section is for those with stable or improved disease following primary systemic therapy. Treatment that focuses on the main tumor site(s) is called local control therapy.

Local control therapy might be:

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

With local control treatment, the goal is to surgically remove all cancer or destroy all cancer cells with RT. Sometimes, surgery with RT is recommended. You will likely need more rounds of systemic therapy after local control treatment. The best treatment for you will be based on the multidisciplinary cancer team recommendations and your wishes.

Monitoring

After completing treatment, you will be monitored for signs that cancer has returned 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.

Disease progression

When cancer does not respond or progresses after primary treatment, then local control therapy such as radiation therapy (RT) 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 4.**

Guide 4
Second-line systemic therapy options: Relapsed, progressed, or metastatic

Preferred options	• Cyclophosphamide and topotecan. Vincristine might be added. • Irinotecan and temozolomide. Vincristine might be added.
Other recommended	• Cabozantinib • Docetaxel and gemcitabine. Vincristine might be added. • Ifosfamide (high-dose) • Regorafenib
Used in some cases	• Ifosfamide, carboplatin, and etoposide. Vincristine might be added. • Lurbinectedin

Metastatic disease

Metastatic Ewing sarcoma is cancer that is 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 than treatment for cancer that has recurred or progressed. Treatment of cancer that has spread or metastasized is explained next.

Local control

Local control therapy focuses on the primary (main) tumor. Options include surgery, radiation therapy (RT), systemic therapy, or a combination of these treatments. Metastasis might be treated separately. Lungs, bone, and bone marrow are the most common sites of metastasis. Often, a lung metastasis is easier to treat than a metastasis found in other sites. Treatment for lung metastasis might include surgery and/or RT.

Widely metastatic

Treatment for widely metastatic disease is either systemic 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 systemic therapy.

Metastatic disease at diagnosis

Systemic therapy is the first or primary treatment for all types of Ewing sarcoma including metastatic disease as the first (initial) diagnosis. Systemic therapy will include a combination of chemotherapies called multi-agent chemotherapy. Systemic therapy options for an initial diagnosis of metastatic Ewing sarcoma can be found in **Guide 5.**

Guide 5
Systemic therapy options: Metastatic disease as initial diagnosis

Preferred options	• Vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide (VDC/IE) • Vincristine, doxorubicin, ifosfamide, and dactinomycin (VAIA) • Vincristine, ifosfamide, doxorubicin, and etoposide (VIDE) • Vincristine, doxorubicin, and cyclophosphamide (VDC)

*Note: Dactinomycin may be substituted for doxorubicin

Key points

- Systemic therapy is the first or primary treatment for Ewing sarcoma. It is a combination of chemotherapies.
- Treatment after first-line systemic therapy is called local control therapy. It focuses on the main tumor site(s). The goal is to remove all cancer with surgery or destroy all cancer cells with radiation therapy. More systemic therapy might be given after surgery or radiation therapy (RT).
- Treatment for an initial diagnosis of metastatic Ewing sarcoma is different than cancer that has spread during or after treatment.
- Metastatic disease at diagnosis is treated with multi-agent chemotherapy. Treatment for cancer that has progressed or spread during or after treatment is often a combination of therapies.

Palliative care is appropriate for anyone, regardless of age, cancer stage, or the need for other therapies. It focuses on physical, emotional, social, and spiritual needs that affect quality of life.

Questions to ask

- Which treatment(s) 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 treatment matter?
- What side effects can I expect from this treatment?

8
Giant cell tumor of bone

Giant cell tumor of bone (GCTB) is usually benign (not cancer). However, it is an aggressive, unpredictable disease that tends to return. Treatment is needed to prevent severe bone damage. Together, you and your care team will choose a treatment plan that is best for you.

Overview

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. When viewed under a microscope, the tumor consists of many very large cells or giant cells. It is not known 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 it can be found in the bones of the arms and the legs. It can also affect flat bones, such as the breastbone or pelvis.

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

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 will have a biopsy to confirm diagnosis and imaging tests to see if the tumor is in one area (localized) or has spread to distant sites (metastasized).

For possible tests before starting treatment, **see Guide 6.**

Guide 6
Possible tests for GCTB

Medical history and physical exam
Imaging of main site
Chest imaging
Bone scan
Biopsy to confirm diagnosis

Localized disease

In localized disease, the tumor is confined to one area. Treatment is based on if the tumor is resectable or unresectable as well as your overall health.

Resectable

A resectable tumor can be removed with surgery. When possible, you will have surgery to remove the tumor. Frequently, surgery involves an intralesional excision or curettage to save normal bone and scrape out all tumor cells. Sometimes, a bigger surgery (wide resection) is required to remove all bone containing the tumor. This wide resection 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.

Unresectable

An unresectable tumor cannot be removed with surgery. This might be based on location or other reasons such as you might have a serious health condition that prevents surgery.

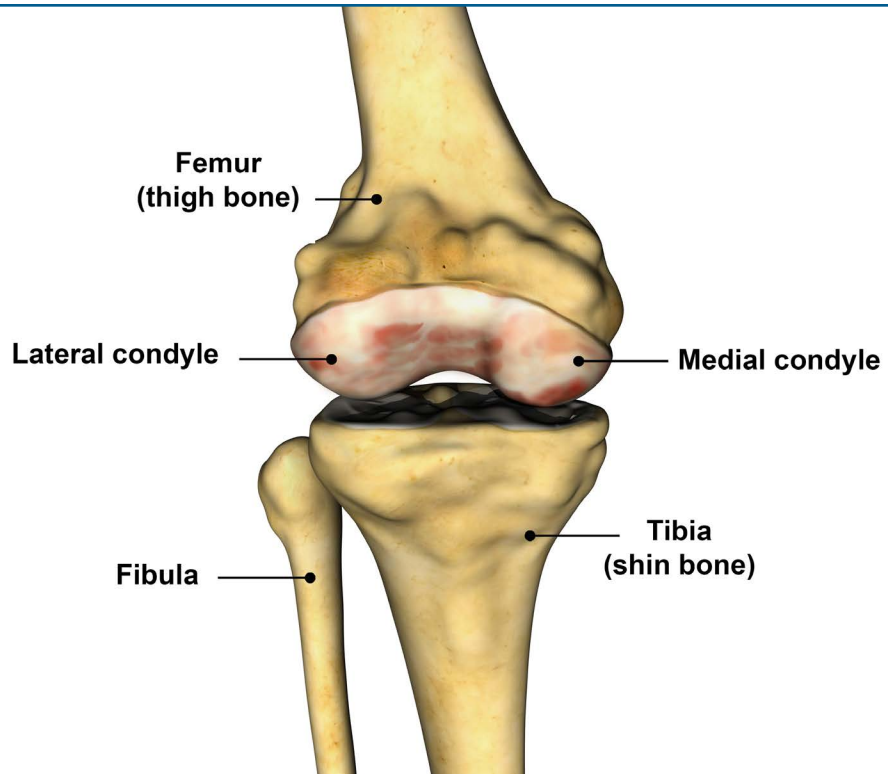
Treatments include:

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

Denosumab (Xgeva) is given to prevent bone loss and fractures. There are some risks with RT and long-term use of denosumab.

Giant cell tumor of bone

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



After treatment you will have imaging tests. If tests find that the tumor is now resectable, you will have surgery to remove the tumor. If the tumor cannot be removed with surgery, you will enter surveillance. Surveillance monitors for any signs that cancer has returned.

Monitoring

After completing treatment, you will be monitored to watch for signs the tumor has returned called recurrence. Monitoring includes imaging tests of the surgical site. You will have chest imaging to look for lung metastases.

Recurrence

GCTB often returns to the same area called local recurrence. There is concern it can spread (metastasize) to the lungs. You should have chest imaging to see if there are any metastases in the lungs. Treatment is based on if it is a local or metastatic recurrence.

Local recurrence

Denosumab might be given before surgery to remove a resectable primary tumor (wide excision). Both resectable and unresectable treatment can be found under *Localized disease* in this chapter.

Metastatic recurrence

For metastatic recurrence, see *Metastatic disease* next.

Denosumab can affect your teeth. Therefore, you should see a dentist before starting denosumab.

Metastatic disease

Metastatic disease at diagnosis is different than malignant transformation. GCTBs are typically non-cancerous. However, in some cases GCTB can transform into a malignant (cancer), more aggressive tumor. If this happens, it will be treated as osteosarcoma, which can be found in the next chapter.

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

- For resectable tumors, you might have surgery to remove the main tumor and metastasis.
- For unresectable tumors, options include denosumab (preferred), radiation therapy, and observation. Observation is sometimes referred to as watch-and-wait. Ask what this might mean for you.

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 will have tests to see if the tumor is in one area (localized) or has spread to distant sites (metastasized). Treatment will be based on if surgery is possible.
- In metastatic disease, there is more than one tumor. GCTB may metastasize to the lungs or other bone.
- After completing treatment, you will be monitored to watch for signs the tumor has returned called recurrence.
- GTCB often returns to the same area called recurrence. Treatment is based on if it 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 treatment matter?
- What side effects can I expect from this treatment?
- What questions should I ask my dentist?

9 Osteosarcoma

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

Overview

Osteosarcoma starts in bone-forming cells called osteoblasts. The cause is not known. Osteosarcoma is commonly found in large bones in the area of bone with the fastest growth rate. However, it can occur in any bone.

Osteosarcoma tends to occur in bones of the:

- Shin (near the knee)
- Thigh (near the knee)
- Upper arm (near the shoulder)

There are 3 main types of osteosarcoma:

- Intramedullary (inside the bone)
- Surface (on the bone surface)
- Extraskeletal (outside the bone, and is a soft tissue sarcoma)

For treating extraskeletal osteosarcomas, see *NCCN Guidelines for Patients®: Soft Tissue Sarcoma* at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.

Testing

Before starting treatment, you will have imaging, biopsy, and blood tests. Treatment can alter fertility. Therefore, you might be referred to a fertility specialist to discuss fertility preservation options before starting treatment. You might also be referred to genetic counseling and genetic testing based on your personal or family history of cancer.

For possible tests, **see Guide 7.**

Guide 7
Possible tests for osteosarcoma

Medical history and physical exam
MRI with or without CT of primary site
Chest imaging including chest CT
FDG-PET/CT and/or bone scan
MRI or CT of metastatic sites
Lactate dehydrogenase (LDH)
Alkaline phosphatase (ALP)
Consider speaking to a fertility specialist before starting treatment
Consider speaking to a genetic counselor if personal or family history of cancer

Systemic therapy

Chemotherapy and other systemic therapies are part of treatment for osteosarcoma. First-line chemotherapies are those given first. Second-line systemic therapy options are used when disease progresses, does not respond to first-line options, or relapses. **See Guide 8 and Guide 9.**

Systemic therapies are often described in the following ways:

- **Preferred therapies** have the most evidence they work better than others.
- **Other recommended therapies** may not work quite as well as preferred therapies, but they can still be useful.
- **Therapies used in certain** cases are for people with specific cancer features or health circumstances.

Guide 8
First-line systemic therapy options

Preferred options	• Cisplatin and doxorubicin • High-dose methotrexate, cisplatin, and doxorubicin (MAP)
Other recommended	• Doxorubicin, cisplatin, ifosfamide, and high-dose methotrexate

Guide 9
Second-line systemic therapy options

Preferred options	• Ifosfamide (high dose) with or without etoposide • Regorafenib • Sorafenib
Other recommended	• Cabozantinib • Cyclophosphamide and topotecan • Gemcitabine with or without docetaxel • Sorafenib with everolimus
Used in some cases	• Cyclophosphamide and etoposide • Ifosfamide, carboplatin, and etoposide • High-dose methotrexate • High-dose methotrexate, etoposide, and ifosfamide • Radium-223 for relapsed or refractory disease beyond second-line therapy

Low-grade and parosteal osteosarcoma

This section is for intramedullary and surface low-grade osteosarcoma. Treatment for low-grade osteosarcoma is surgery to remove the tumor called wide excision. Systemic therapy might follow surgery if a high-grade component is found unexpectedly in the surgical specimen. Parosteal osteosarcoma usually only requires surgery. Dedifferentiated parosteal osteosarcomas are not considered to be low-grade tumors.

High-grade osteosarcoma

This section is for intramedullary and surface high-grade osteosarcoma. Other types of high-grade non-osteosarcoma such as undifferentiated pleomorphic sarcoma (UPS) of bone could also be treated the same way.

Systemic therapy before surgery

For most, treatment starts with preoperative chemotherapy. The goal of chemotherapy is to kill the cancer cells before surgery. After chemotherapy, you will have imaging tests to restage your cancer, assess treatment response, and plan surgery. For preoperative chemotherapy options, **see Guide 8.**

Surgery or local control therapy

Most osteosarcomas are treated with surgery to remove all of the cancer cells in the body. Imaging scans help determine if the osteosarcoma can be removed with surgery. Most osteosarcomas are resectable with limb-sparing surgery (LSS). This means the surgeon removes the bone containing cancer and reconstructs the limb. Some osteosarcomas require amputation. Rarely, the tumor is unresectable. In this case, treatment then involves radiation therapy or systemic therapy.

Systemic therapy after surgery

Treatment after surgery is called adjuvant treatment. When no cancer is found in the surgical margin, it is called a negative margin. Treatment is systemic therapy. When cancer remains in the surgical margin, it is called a positive margin. Treatment might be systemic therapy, or more surgery. Radiation therapy might follow surgery.

Metastatic disease at diagnosis

This section is for those with metastatic disease at diagnosis. Metastases can be found in the lungs (pulmonary), bone (skeletal), or internal organs (visceral). These are often resectable. Surgery to remove a metastasis is called a metastasectomy. This is different from a mastectomy, which is surgery to remove the breast. Some metastases might be treated with systemic therapy, radiation therapy, or other therapies. For example, unresectable lung metastases might be treated with ablation.

If surgery is not possible, then treatment might be systemic therapy or radiation therapy. Before starting systemic therapy, biomarker testing might be done.

Periosteal osteosarcoma

Periosteal osteosarcoma starts on the surface of the bone called the periosteum. Treatment is surgery to remove the tumor called wide excision. Systemic therapy might be given before surgery to reduce the size of tumor or the amount of cancer.

Monitoring

After completing treatment, you will be monitored to watch for signs that cancer has returned called recurrence or relapse. Monitoring includes physical exam, blood and imaging tests, functional movement assessment, and regular visits with your orthopedic surgeon and oncologist.

Relapse

If cancer returns, treatment is systemic therapy and/or surgery. Surgery is not always possible. Systemic therapy options can be found in **Guide 9.**

After treatment for relapse, you will have imaging tests. If your cancer has responded to treatment, then you will be monitored for the return of cancer (relapse) or for progression.

Treatment options for relapse and disease progression include:

- Surgery to remove tumor (excision), if possible
- Clinical trial
- Radiation therapy (may include radiopharmaceuticals)
- Best supportive care

Standard of care is the best-known way to treat a particular disease based on past clinical trials. There may be more than one treatment regimen that is considered standard of care. Ask your care team what treatment options are available and if a clinical trial might be right for you.

Key points

- Osteosarcoma is the most common type of bone cancer. It usually occurs around the knee or shoulder.
- Treatment most commonly involves systemic therapy and surgery. Parosteal osteosarcoma usually only requires surgery.
- Osteosarcoma can metastasize in the lungs (pulmonary), bone (skeletal), or internal organs (visceral). These are often resectable.
- Best supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life.
- A metastasectomy is surgery to remove a metastasis.
- After completing treatment, you will be monitored to watch for signs that cancer has returned called recurrence or relapse. This is called surveillance.

Questions to ask

- Which option is proven to work best for my type of cancer, age, overall health, and other factors?
- What are all of 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?

10
Other resources

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. It plainly explains expert recommendations and suggests questions to ask your care team. 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. They often have 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 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
Imermanangels.org

MIB Agents Osteosarcoma Alliance
Mibagents.org

MedlinePlus
medlineplus.gov

National Cancer Institute (NCI)
cancer.gov/types

National Coalition for Cancer Survivorship
canceradvocacy.org

Northwest Sarcoma Foundation
nwsarcoma.org

The Alan B. Slifka Foundation
slifkafoundation.org

The Paula Takacs Foundation for Sarcoma Research
paulatakacsfoundation.org

Triage Cancer
triagecancer.org

Questions to ask about resources and support

- 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?

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

Words to know

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

biomarker testing
A lab test of any molecule in your body that can be measured to assess your health. 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.

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 (CBC)
A lab test that includes the number of blood cells.

computed tomography (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.

curettage
Surgery to remove part of the tumor. Also called intralesional excision.

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

external beam radiation therapy (EBRT)
A cancer treatment with radiation received from a machine outside the body.

functional assessment
A test that looks at your ability to manage tasks and activities that are used in daily life.

hereditary
Passed down from birth parent to child through coded information in cells (genes).

histology
The study of tissues and cells under a microscope.

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 much cancer cells look like normal cells.

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

interventional radiologist
A doctor who is an expert in imaging tests and using image-guided tools to perform minimally invasive techniques to diagnose or treat disease.

intralesional excision
Surgery to remove part of the tumor. Also called curettage.

magnetic resonance imaging (MRI)
A test that uses radio waves and powerful magnets to make pictures of the insides of the body.

medical oncologist
A doctor who is an expert in cancer drugs.

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

metastasectomy
Surgery to remove a metastasis.

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.

ossification
Process of new bone formation.

osteoblasts
Bone-forming cells.

osteoclasts
Bone-dissolving cells.

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

pathologist
A doctor who is an expert in testing cells and tissue to find disease.

positron emission tomography (PET)
A test that uses radioactive material to see the shape and function of body parts.

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

primary tumor
The first mass of cancerous cells.

prognosis
The pattern and outcome of a disease.

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

radiation therapy (RT)
A treatment that uses high-energy rays.

radiologist
A doctor who is an expert in imaging tests.

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.

remodeling
Process where bone cells are resorbed and new cells are formed.

resectable
A tumor that can be removed completely with surgery.

sarcoma
A cancer of bone or soft tissue cells.

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 palliative care or best supportive care.

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

systemic therapy
Treatment that works throughout the body.

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

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

unresectable
A tumor that cannot be removed with surgery.

wide excision
Surgery to remove all of the tumor.

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

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This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Bone Cancer, Version 1.2025. It was adapted, reviewed, and published with help from the following people:

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NCCN Cancer Centers

Abramson Cancer Center
at the University of Pennsylvania
Philadelphia, Pennsylvania
800.789.7366 • pennmedicine.org/cancer

Case Comprehensive Cancer Center/
University Hospitals Seidman Cancer Center and
Cleveland Clinic Taussig Cancer Institute
Cleveland, Ohio
UH Seidman Cancer Center
800.641.2422 • uhhospitals.org/services/cancer-services
CC Taussig Cancer Institute
866.223.8100 • my.clevelandclinic.org/departments/cancer
Case CCC
216.844.8797 • case.edu/cancer

City of Hope National Medical Center
Duarte, California
800.826.4673 • cityofhope.org

Dana-Farber/Brigham and Women's Cancer Center |
Mass General Cancer Center
Boston, Massachusetts
877.442.3324 • youhaveus.org
617.726.5130 • massgeneral.org/cancer-center

Duke Cancer Institute
Durham, North Carolina
888.275.3853 • dukecancerinstitute.org

Fox Chase Cancer Center
Philadelphia, Pennsylvania
888.369.2427 • foxchase.org

Fred & Pamela Buffett Cancer Center
Omaha, Nebraska
402.559.5600 • unmc.edu/cancercenter

Fred Hutchinson Cancer Center
Seattle, Washington
206.667.5000 • fredhutch.org

Huntsman Cancer Institute at the University of Utah
Salt Lake City, Utah
800.824.2073 • healthcare.utah.edu/huntsmancancerinstitute

Indiana University Melvin and Bren Simon
Comprehensive Cancer Center
Indianapolis, Indiana
888.600.4822 • www.cancer.iu.edu

Johns Hopkins Kimmel Cancer Center
Baltimore, Maryland
410.955.8964
www.hopkinskimmelcancercenter.org

Mayo Clinic Comprehensive Cancer Center
Phoenix/Scottsdale, Arizona
Jacksonville, Florida
Rochester, Minnesota
480.301.8000 • Arizona
904.953.0853 • Florida
507.538.3270 • Minnesota
mayoclinic.org/cancercenter

Memorial Sloan Kettering Cancer Center
New York, New York
800.525.2225 • mskcc.org

Moffitt Cancer Center
Tampa, Florida
888.663.3488 • moffitt.org

O'Neal Comprehensive Cancer Center at UAB
Birmingham, Alabama
800.822.0933 • uab.edu/onealcancercenter

Robert H. Lurie Comprehensive Cancer Center
of Northwestern University
Chicago, Illinois
866.587.4322 • cancer.northwestern.edu

Roswell Park Comprehensive Cancer Center
Buffalo, New York
877.275.7724 • roswellpark.org

Siteman Cancer Center at Barnes-Jewish Hospital
and Washington University School of Medicine
St. Louis, Missouri
800.600.3606 • siteman.wustl.edu

St. Jude Children's Research Hospital/
The University of Tennessee Health Science Center
Memphis, Tennessee
866.278.5833 • stjude.org
901.448.5500 • uthsc.edu

Stanford Cancer Institute
Stanford, California
877.668.7535 • cancer.stanford.edu

The Ohio State University Comprehensive Cancer Center -
James Cancer Hospital and Solove Research Institute
Columbus, Ohio
800.293.5066 • cancer.osu.edu

The UChicago Medicine Comprehensive Cancer Center
Chicago, Illinois
773.702.1000 • uchicagomedicine.org/cancer

The University of Texas MD Anderson Cancer Center
Houston, Texas
844.269.5922 • mdanderson.org

UC Davis Comprehensive Cancer Center
Sacramento, California
916.734.5959 • 800.770.9261
health.ucdavis.edu/cancer

UC San Diego Moores Cancer Center
La Jolla, California
858.822.6100 • cancer.ucsd.edu

UCLA Jonsson Comprehensive Cancer Center
Los Angeles, California
310.825.5268 • uclahealth.org/cancer

UCSF Helen Diller Family Comprehensive Cancer Center
San Francisco, California
800.689.8273 • cancer.ucsf.edu

University of Colorado Cancer Center
Aurora, Colorado
720.848.0300 • coloradocancercenter.org

University of Michigan Rogel Cancer Center
Ann Arbor, Michigan
800.865.1125 • rogelcancercenter.org

University of Wisconsin Carbone Cancer Center
Madison, Wisconsin
608.265.1700 • uwhealth.org/cancer

UT Southwestern Simmons Comprehensive Cancer Center
Dallas, Texas
214.648.3111 • utsouthwestern.edu/simmons

Vanderbilt-Ingram Cancer Center
Nashville, Tennessee
877.936.8422 • vicc.org

Yale Cancer Center/Smilow Cancer Hospital
New Haven, Connecticut
855.4.SMILOW • yalecancercenter.org

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