



NCCN
GUIDELINES
FOR PATIENTS®

2023

Bone Cancer



Presented with support from



NATIONAL COMPREHENSIVE CANCER NETWORK®
FOUNDATION
Guiding Treatment. Changing Lives.

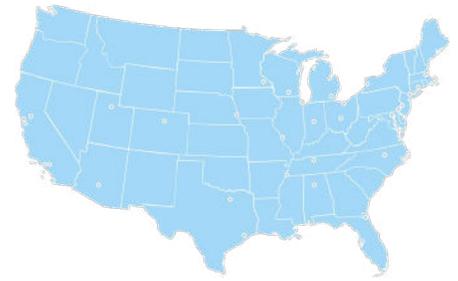
Available online at
[NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines)



About the NCCN Guidelines for Patients®



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 2.2023 – September 28, 2022.

View the NCCN Guidelines
for Patients free online
[NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines)

Find an NCCN Cancer
Center near you
[NCCN.org/cancercenters](https://www.nccn.org/cancercenters)

Connect with us     YouTube 

Supporters



NCCN Guidelines for Patients are supported by funding from the NCCN Foundation®

NCCN Foundation gratefully acknowledges the following corporate supporters for helping to make available these NCCN Guidelines for Patients: Amgen Inc.

NCCN independently adapts, updates, and hosts the NCCN Guidelines for Patients. Our corporate supporters do not participate in the development of the NCCN Guidelines for Patients and are not responsible for the content and recommendations contained therein.

Additional support is provided by



Founded in 1965, the Alan B. Slifka Foundation is a private family foundation dedicated to the pursuit of inclusion, diversity, peace and healing. Among the Foundation's priorities is funding research into less toxic, more effective, and potentially curative therapies for a variety of cancers but especially for connective tissue cancers, known as sarcomas, that disproportionately impact children and young adults. slifkafoundation.org

To make a gift or learn more visit online or email

NCCNFoundation.org/donate

PatientGuidelines@NCCN.org

Contents

- 4 Bone cancer basics
- 9 Testing for bone cancer
- 21 Cancer staging
- 28 Treatment overview
- 38 Chondrosarcoma
- 42 Chordoma
- 47 Ewing sarcoma
- 53 Giant cell tumor of bone
- 57 Osteosarcoma
- 63 Making treatment decisions
- 77 Words to know
- 80 NCCN Contributors
- 81 NCCN Cancer Centers
- 82 Index

© 2022 National Comprehensive Cancer Network, Inc. All rights reserved. NCCN Guidelines for Patients and illustrations herein may not be reproduced in any form for any purpose without the express written permission of NCCN. No one, including doctors or patients, may use the NCCN Guidelines for Patients for any commercial purpose and may not claim, represent, or imply that the NCCN Guidelines for Patients that have been modified in any manner are derived from, based on, related to, or arise out of the NCCN Guidelines for Patients. The NCCN Guidelines are a work in progress that may be redefined as often as new significant data become available. NCCN makes no warranties of any kind whatsoever regarding its content, use, or application and disclaims any responsibility for its application or use in any way.

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](https://www.nccn.org/patients).

National Comprehensive Cancer Network (NCCN) and NCCN Foundation
3025 Chemical Road, Suite 100, Plymouth Meeting, PA 19462 USA

1

Bone cancer basics

- 5 The bone
- 6 How bone is formed
- 7 Types of bone tumors
- 7 Bone cancer
- 8 How bone cancer spreads
- 8 Key points

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 or metastasize to the bone. This chapter presents an overview of bone cancer.

The bone

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

Our body has about 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.

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
- Spongy 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.

The skeletal system

The human skeletal system consists of bones, cartilage, ligaments, and tendons. It includes all of the bones and joints in the body.



How bone is 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.

There are 2 forms of ossification:

- Compact bone (endochondral), which makes up most of the skeleton
- Cancellous bone (intermembranous), which includes parts of the skull, the shoulder blades, and the ends of the long bones. Long bones are longer than they are wide.

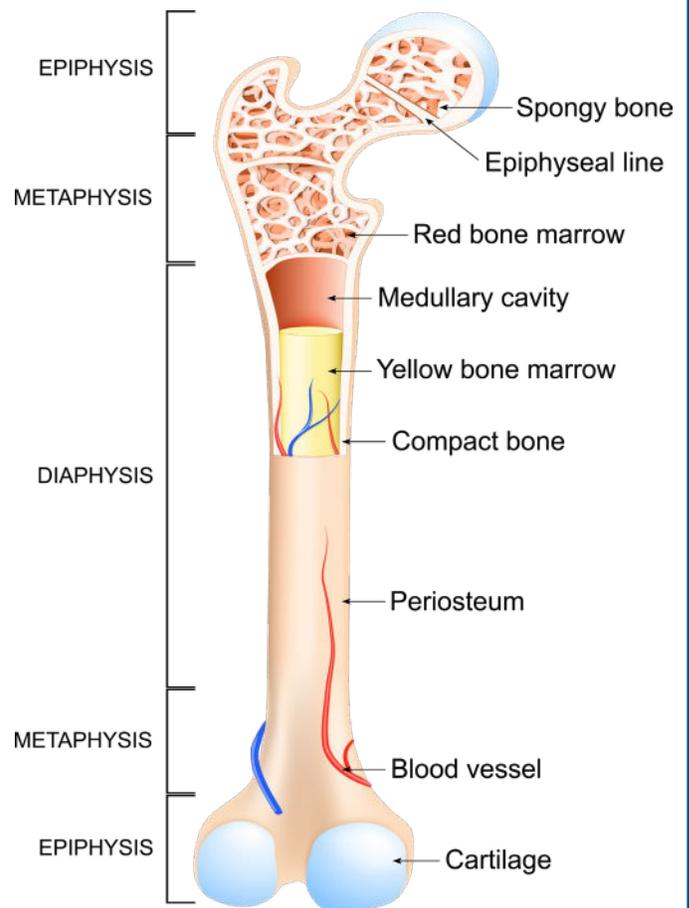
Once ossification is complete, bone is continuously being replaced by a process called remodeling. Remodeling breaks down and rebuilds bone and helps maintain normal calcium levels in the body. When calcium is needed, cells called osteoclasts break down bone. This releases calcium into the bloodstream. 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 physal scar or line visible in the bone

Anatomy of the bone

The epiphysis is the end of the bone. The metaphysis is located between the physis (growth plate) and the diaphysis. The diaphysis is the middle region of the bone.



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). The cause of bone tumors is unknown. They 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 epiphysis.
- **Osteosarcoma** – starts in cells that form bone in the metaphysis near the growth plates.

Since 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 treatment center or hospital that has both experts and experience in your type of bone cancer.

Bone cancer

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

Sarcomas

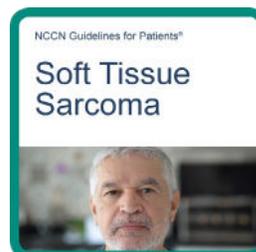
Most primary bone cancers are sarcomas. Sarcomas are a rare group of cancers.

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.

More information on soft tissue sarcomas can be found at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.



How bone cancer spreads

Unlike normal cells, cancer cells can grow or spread to form tumors in other parts of the body. This is called a metastasis.

Cancer can spread through blood or the lymphatic system. The lymphatic system contains a clear fluid called lymph. Lymph gives cells water and food and contains white blood cells that fight germs. Lymph nodes filter lymph and remove the germs. Lymph travels throughout the body in vessels like blood does. Lymph vessels and nodes are found everywhere in the body. Although cancer can spread through your blood, you cannot spread your cancer to other people if they are exposed to your blood.

Bone cancer can spread to the lung or other bones. Bone cancer that has metastasized to the lung is still called bone cancer.

Key points

- ▶ Bone cancer is a group of cancers that start in the bone or cartilage. This is called primary bone cancer. Bone cancers that start in another part of the body (such as the breast, lungs, or kidney) are called secondary or metastatic bone tumors.
- ▶ Bone tumors may be cancerous (malignant) or noncancerous (benign). The cause of bone tumors is unknown.
- ▶ Bone sarcomas, such as osteosarcomas, start in the cells that form bone. Most primary bone cancers are sarcomas.
- ▶ Osteosarcoma, Ewing sarcoma, and chondrosarcoma are the most common types of bone cancer.
- ▶ Bone cancer can metastasize to the lungs and other bones.
- ▶ It is important to find a treatment center or hospital that has both experts and experience in your type of bone cancer.

2

Testing for bone cancer

- 10 Test results
- 10 General health tests
- 12 Fertility (all genders)
- 12 Preventing pregnancy
- 13 Performance status
- 13 Blood tests
- 15 Imaging tests
- 17 Biopsy
- 17 Genetic testing
- 18 Biomarker tests
- 20 Key points

Treatment planning starts with testing. Accurate testing is needed to diagnose and treat primary bone cancer. A biopsy is recommended before starting treatment. This chapter presents an overview of the tests you might receive and what to expect.

Test results

Results from imaging studies and biopsies will be used to determine your treatment plan. It is important you understand what these tests mean. Ask questions and keep copies of your test results. Online patient portals are a great way to access your test results.

Keep these things in mind:

- Choose a friend, family member, or peer who can drive you to appointments, provide meals, or offer emotional support during diagnosis and treatment.
- Bring someone with you to doctor visits, if possible.
- Write down questions and take notes during appointments. Don't be afraid to ask your care team questions. Get to know your care team and help them get to know you.
- Get copies of blood tests, imaging results, and reports about the specific type of cancer you have.

- Bring all images on a compact disc (CD). You can request a disc from the hospital or center where you had your imaging. X-rays, MRIs, and CT scans cannot be faxed. If you are meeting a new doctor, please bring all your images on a CD.
- Organize your papers. Create files for insurance forms, medical records, and test results. You can do the same on your computer.
- Keep a list of contact information for everyone on your care team. Add it to your phone. Hang the list on your refrigerator or keep it in a place where someone can access it in an emergency. Keep your primary care physician (PCP) informed of changes to this list. You are encouraged to keep your PCP. They are great partners in your care.
- Include in your contact list information on the exact type of cancer, as well as any treatment and the date it started.

General health tests

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 medicines, herbals, or supplements you take. Some supplements interact and affect prescriptions that your doctor may give you. Tell your doctor about any symptoms you have. A medical history, sometimes called a health history, will help determine which treatment is best for you.

Family history

Some cancers and other diseases can run in families. Your doctor 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, and if it is in multiple locations.

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.

Bring a list of any medications, vitamins, over-the-counter drugs, herbals, or supplements you are taking.

For possible tests, **see Guide 1**.

Guide 1

Possible tests: Those 40 years of age and over

Medical history and physical exam

Bone scan

Chest x-ray

Blood tests such as complete blood count (CBC), comprehensive metabolic panel (CMP) with calcium to assess for hypercalcemia

Chest/abdominal/pelvic CT with contrast

Biopsy, as needed, performed by orthopedic oncologist at center where you will be treated

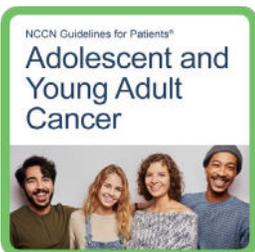
* Those under 40 years of age will be referred to an orthopedic oncologist

Fertility (all genders)

Treatment such as chemotherapy 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 really sure at the moment. Fertility and reproductive specialists can help you sort through what may be best for your situation.

More information on fertility preservation in adolescents and young adults can be found at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.



Changes in fertility

Treatment might cause your fertility to be temporarily impaired or interrupted. This temporary 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

Preventing pregnancy during treatment is important. Cancer and cancer treatment can affect the ovaries and damage sperm. 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.

Those with ovaries

Those who can become pregnant will have a pregnancy test before starting treatment. Cancer treatment can hurt the developing baby if you are or become pregnant during treatment. Therefore, birth control to prevent pregnancy during and after treatment is recommended. If you are pregnant or breastfeeding at the time of your cancer diagnosis, certain treatments will need to be avoided.

Menstruation, menses, menstrual flow, or your “period” may stop during treatment, but often returns within 2 years after treatment in those 40 years of age and under. It is still possible to become pregnant even though you might not have a period. Therefore, birth control is recommended during and after treatment. Consult your doctor for the best time to plan a pregnancy.

Those with testicles

Cancer and cancer treatment can damage sperm. Therefore, use contraception (birth control) such as condoms to prevent pregnancy during and immediately after cancer treatment.

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 ECOG (Eastern Cooperative Oncology Group). PS is one factor taken into consideration when choosing a treatment plan. Your preferences about treatment are always important.

The ECOG PS scores range from 0 to 5.

- **PS 0** means the person is fully active.
- **PS 1** means the person is still able to perform light to moderate activity, but with some limitations.
- **PS 2** means the person is limited to the chair or bed less than half of the time and still able to care for self.
- **PS 3** means the person is limited to the chair or bed more than half of the time.
- **PS 4** means the person is totally confined to the bed or chair and completely unable to care for self.
- **PS 5** means the person is not alive.

Good PS is usually PS 0 or PS 1.

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 your vein. Some blood tests you might have are described next.

Alkaline phosphatase

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

Performance status (PS) is a person's general level of fitness and ability to perform daily tasks.



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. Red blood cells carry oxygen throughout your body, white blood cells fight infection, and platelets control bleeding.

Comprehensive metabolic panel

A comprehensive metabolic panel (CMP) measures 14 different substances in your blood. It is usually done on the plasma part of your blood. A CMP 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 the most common mineral in the body. It is needed for healthy teeth, bones, and other body tissues. Bone damage from cancer can cause your bones to release calcium into the bloodstream.

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 will be given a pregnancy test before treatment begins.

Those with bone cancer should be treated at centers experienced in this type of cancer.

Imaging tests

Imaging tests take pictures of the inside of your body. Imaging tests show the primary tumor, or where the cancer started, and look for cancer in other parts of the body. A radiologist, who is an expert in interpreting imaging tests, will write a report and send this report to your doctor. Your doctor will discuss the results with you.

You will 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. However, the test is quick and painless.

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.

CT scan

A computed tomography (CT or CAT) 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. Intravenous (IV) contrast is often used.

CT machine

A CT machine is large and has a tunnel in the middle. During the test, you will lie on the table that moves slowly through the tunnel.



Contrast

Contrast material is used to improve the pictures of the inside of the body. Contrast materials are not dyes, but 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 the body in your urine immediately after the test. The types of contrast vary and are different for CT and MRI.

Tell your doctors if you have had allergic reactions to contrast in the past, especially to iodine or to shellfish such as shrimp. This is important. You might be given medicines, such as Benadryl and prednisone, 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.

Mammogram

A mammogram is a picture of the insides of your breast. The pictures are made using x-rays. A computer combines the x-rays to make detailed pictures. Diagnostic mammograms are made with more x-rays than screening mammograms. A bilateral mammogram includes pictures of both breasts. It is used to identify any tumors, and if so, the number and size of the tumor(s). A mammogram might be needed to confirm that the cancer found in your bones is not from breast cancer.

MRI scan

A magnetic resonance imaging (MRI) scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays. During the test, you will likely be asked to hold your breath for 10 to 20 seconds as the technician collects the images. Contrast might be used.

A closed MRI has a capsule-like design where the magnet surrounds you. An open MRI has a magnet top and bottom, which allows for an opening on each end. Most people prefer to find a location with an open MRI. Tell your doctor if you have any metal in your body or if you tend to have claustrophobia (a dread or fear of enclosed spaces). You might be surprised how tight a fit it is and how confined you might feel even in an open MRI.

PET/CT scan

A positron emission tomography (PET) 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 if they are using sugar produced by your body to grow. 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 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. It may be done with one or two machines depending on the cancer center. A whole-body PET/CT or head-to-toe scan can be used to look for evidence of cancer throughout your body.

Biopsy

A biopsy is the removal of a sample of tissue 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 at an experienced center where you will receive treatment.

- **Core needle biopsy** removes tissue samples with a large, 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 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

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 testing

Genetic testing is done using blood or saliva (spitting into a cup). The goal is to look for gene mutations inherited from your biological 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, family members might carry these mutations. Tell your doctor if there is a family history of cancer.

There are 3 major types of genetic testing:

- **Cytogenetic** – to examine whole chromosomes
- **Biochemical** – to measure proteins produced by genes
- **Molecular** – to look for DNA or gene mutations and/or gene fusions

While it can be confusing, just know that testing done to look for an inherited gene mutation or an inherited risk of cancer is different than genetic testing done on cancer cells or testing to look for proteins produced by cancer cells.

Biomarker tests

Inside our cells are deoxyribonucleic acid (DNA) molecules. These molecules are tightly packaged into what is called a chromosome. Chromosomes contain most of the genetic information in a cell. Normal human cells contain 23 pairs of chromosomes for a total of 46 chromosomes. Each chromosome contains thousands of genes. Genes are coded instructions for the proteins your cells make. A mutation is when something goes wrong in the genetic code.

A sample from your biopsy will undergo lab tests to look for specific DNA mutations/alterations, protein levels, or other molecular features. This information is used to learn more about your cancer and to choose the best treatment for you. It is sometimes called molecular testing, tumor profiling, tumor sequencing, gene expression profiling, or genomic testing.

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 chromosome 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 parents, but happens for some other reason. When this occurs only in cancer cells, 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 for 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 an option for those with unresectable or metastatic, microsatellite instability-high (MSI-H) solid tumors that have progressed following prior treatment.

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 in some cases for TMB-H tumors.

Tumor mutation testing

A sample of your tumor or blood may be used to see if the cancer cells have any specific mutations. In tumor mutation testing, only the tumor is tested and not the rest of your body.

IDH1 mutation

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

Next-generation sequencing

Next-generation sequencing (NGS) is a high-throughput method used to determine the DNA sequence or gene changes of cancer cells in your bone tumor.

PCR

A polymerase chain reaction (PCR) is a lab process that can make millions or billions of copies of your DNA (genetic information). PCR is very sensitive. It can find 1 abnormal cell among more than 100,000 normal cells. These copies are called PCR product.

What is your family health history?

Some cancers and other diseases run in families—those who are related to you through genes passed down from parent to child. This information is called a family health history. You can ask family members about their health issues like heart disease, cancer, and diabetes, and at what age they were diagnosed. For relatives who have died, ask about the cause and age of death.

Start by asking your parents, siblings, and children. Next, talk to half-siblings, aunts and uncles, nieces and nephews, grandparents, and grandchildren.

Write down what you learn about your family history and share this information with your health care provider.

Some of the questions to ask include:

- ✓ Do you have any chronic diseases, such as heart disease or diabetes, or health conditions such as high blood pressure or high cholesterol?
- ✓ Have you had any other diseases, such as cancer or stroke?
- ✓ How old were you when each of these diseases and health conditions was diagnosed?
- ✓ What is our family's ancestry—from what countries did our ancestors originate?

Key points

- Tests are used to plan treatment and check how well treatment is working.
- Online portals are a great way to access your test results.
- Bone cancer survivors are at risk for fertility issues. Those who want to have children in the future should be referred to a fertility specialist before starting treatment to discuss the options.
- Blood, imaging, and tissue tests check for signs of disease.
- 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.



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.”

3

Cancer staging

- 22 Overview
- 22 TNM score
- 27 Numbered stages
- 27 Key points

Cancer staging is used to 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 cell type (such as sarcoma or carcinoma)
- 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 and how likely the tumor is to grow and spread

TNM score

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

T = Tumor

The primary 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 are staged based on location.

Tumors in limbs, torso, skull, and face

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.

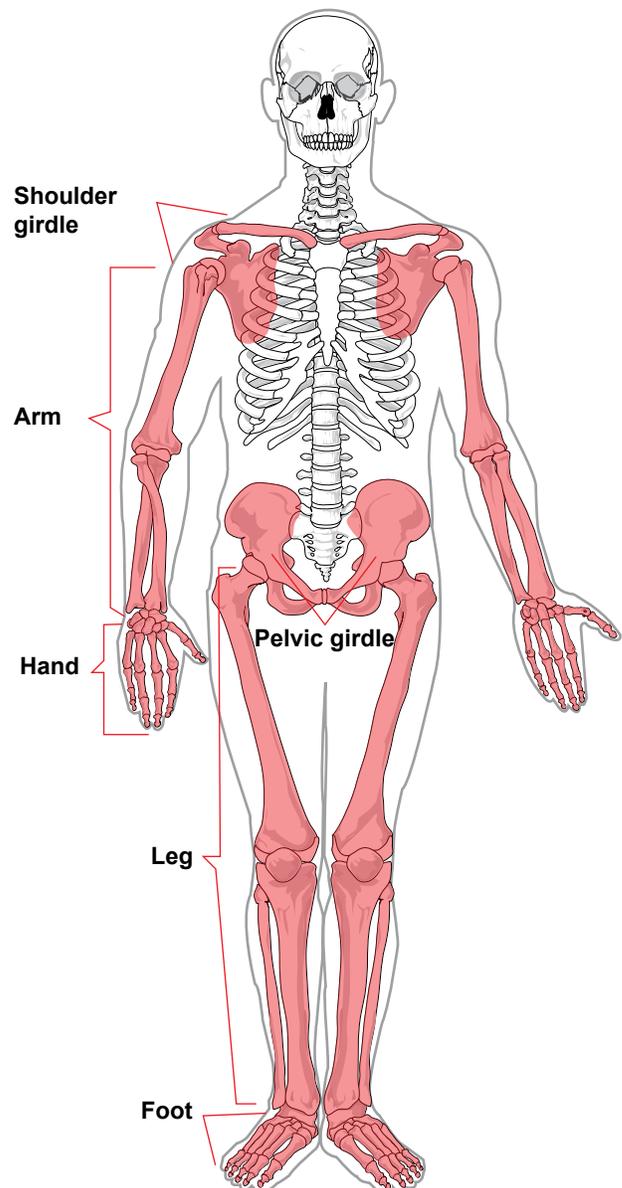
Staging for tumors found in the appendicular skeleton, torso, skull, and face are described next.

- **TX** – Primary (main) 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

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.

Reproduced with permission by Cancer Research UK / Wikimedia Commons.



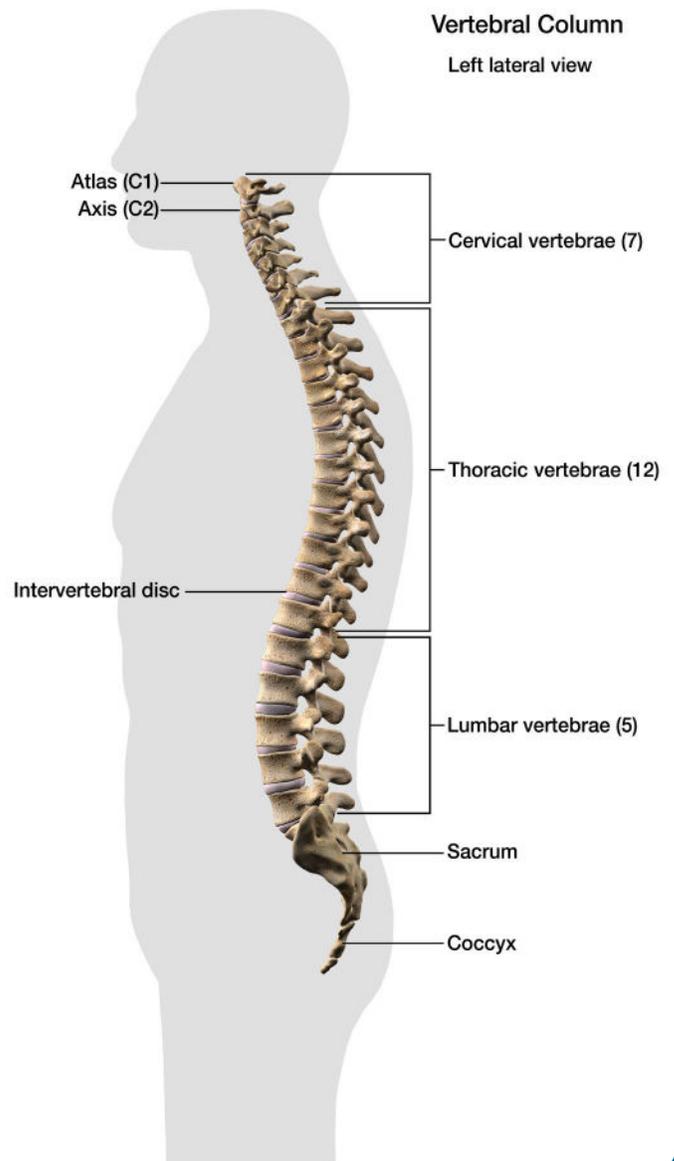
Tumors in spine

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 adjacent vertebrae
- **T2** – Tumor confined to 3 adjacent vertebrae
- **T3** – Tumor confined to 4 or more adjacent vertebrae, or any vertebrae not next to one another
- **T4** – Tumor has grown into spinal canal or large (great) vessels

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).



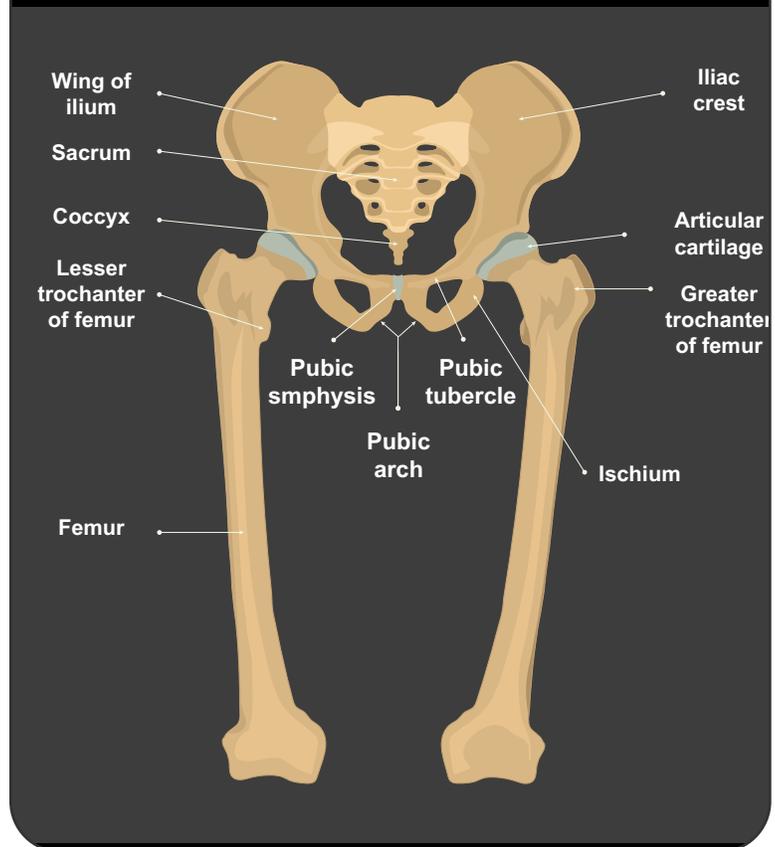
Tumors in pelvis

Staging for tumors found in the pelvis are described next.

- **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 is spanning two pelvic segments but 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 sacroiliac joint and extends into the spine
 - **T4b** – Tumor has grown into major blood vessels

The pelvis

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



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

Cancer that has spread to distant parts of the body is shown as M1. When cancer is not metastatic it is shown as M0. Common sites for metastasis include the lung and bone.

- **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

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)



We want your feedback!

Our goal is to provide helpful and easy-to-understand information on cancer.

Take our [survey](#) to let us know what we got right and what we could do better.

[NCCN.org/patients/feedback](https://www.nccn.org/patients/feedback)

Numbered stages

Number stages range from stage 1 to stage 4, with 4 being the most advanced. These stages are written as stage I, stage II, stage III, and stage IV. Not all bone cancers are described this way. **See Guide 2.**

Guide 2

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 metastatic cancer.
- Common sites for metastasis include the lung and bone.

4

Treatment overview

- 29 Care team
- 30 Surgery
- 32 Systemic therapy
- 33 Radiation therapy
- 34 Clinical trials
- 36 Other treatments
- 37 Key points

Treatment for bone cancer is based on the type, size, and location of the tumor. This chapter presents an overview of the types of treatment and what to expect. Not everyone will receive the same treatment.

Care team

Treating bone cancer takes a team approach. Treatment decisions should involve a multidisciplinary team (MDT). An MDT is a team of doctors, health care workers, and social care professionals from different professional backgrounds who have knowledge (expertise) and experience with 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.

The MDT should meet on a regular basis and should include the following:

- **An orthopedic oncologist** 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:

- **A thoracic surgeon** who performs operations within the chest.
- **A plastic surgeon** who performs operations to improve function or reconstruct missing areas.
- **An interventional radiologist** who performs needle biopsies, ablation therapies, and embolizations, and places ports for treatment.
- **A physiatrist**, a medical doctor who specializes in physical movement and rehabilitation.
- **A vascular surgeon** who specializes in procedures involving arteries, veins, and lymph circulation.
- **A surgical oncologist or general surgeon** who has specialized knowledge and experience related to the diagnosis, preoperative, operative, and postoperative care for the whole patient.
- **A neurosurgeon** who specializes in surgery on the nervous system, especially the brain and spinal cord.

- **An orthopedic oncologist or orthopedic surgeon** who focuses on the body's musculoskeletal system, which includes bones, joints, muscles, ligaments, tendons, and nerves.
- **An orthopedic spine surgeon** who specializes in the diagnosis and treatment of spinal diseases and conditions. May be an orthopedic oncologist.

Your physical, mental, and emotional well-being are important. You know yourself better than anyone. Help other team members understand:

- How you feel
- What you need
- What is working and what is not

Keep a list of names and contact information for each member of your team. This will make it easier for you and anyone involved in your care to know who to contact with questions or concerns.

Surgery

Surgery is an operation or procedure to remove cancer from the body. The type of surgery depends on size, location, and number of tumor(s).

Surgery can be used to:

- Remove all of the tumor
- 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.

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 resection

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. A surgeon who is an expert in your type of bone cancer should be consulted before deciding upon surgical treatment options. Rehabilitation, such as physical and occupational therapy, will likely be part of a limb-sparing treatment plan. This 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.

Amputation

Amputation is the removal of a limb or other body part. A surgeon who is an expert in your type of bone cancer should be consulted before amputation. One type of amputation is a rotationplasty. In a rotationplasty, the bottom of the femur, the knee, and the top of the tibia are removed. The lower leg is then rotated and attached to the femur. The intention of this surgery is to allow knee function when it otherwise would not be possible.

Rehabilitation, such as physical and occupational therapy, will be part of any amputation. 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. A physiatrist is a medical doctor who specializes in rehabilitation.

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.

Order of treatments

Most people with cancer will receive more than one type of treatment. Below is an overview of the order of treatments.

- ✓ **Preoperative or neoadjuvant (before) therapy** is given to shrink the tumor before a primary treatment such as surgery.
- ✓ **Perioperative therapy** is systemic therapy, such as chemotherapy, given before and after surgery.
- ✓ **Primary treatment** is the main treatment given to rid the body of cancer.
- ✓ **Postoperative or adjuvant (after) therapy** is given after primary treatment to rid the body of any cancer cells left behind from surgery. It is also used when the risk of cancer returning (recurrence) is felt to be high.
- ✓ **Definitive treatment** is the best treatment after all choices have been considered.
- ✓ **First-line therapy** is the first set of systemic (drug) treatment given.
- ✓ **Second-line therapy** is the next set of treatment given if cancer progresses during or after systemic therapy.

Talk to your care team about your treatment plan and what it means for your stage and type of cancer.

Systemic therapy

Systemic therapy is drug therapy that works throughout the body. Each works differently. 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. The choice of therapy takes into consideration many factors, including age, other serious health issues, and future treatment possibilities. Your preferences about treatment are important. If you have any religious or personal beliefs about certain kinds of treatment, now would be the time to share them with your care team.

Warnings!

You might be asked to stop taking or avoid certain food or herbal supplements when on a systemic therapy. Some supplements and food 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
- Gingko biloba
- Green tea extract
- St. John's Wort

Certain medicines can also affect the ability of a drug to do its job. Antacids, heart medicine, and antidepressants are just some of the medicines that might interact with a systemic therapy. This is why it is important to tell your doctor about any medications, vitamins, over-the-counter (OTC) drugs, herbals, or supplements you are taking. **Bring a list with you to every visit.**

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 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 will have tests to see how well treatment is working. You might spend time in the hospital during treatment.

Targeted therapy

Targeted therapy is drug 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 (first) 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 contain a radioactive substance that emits radiation. This radioactive substance is different than contrast material used in imaging. Samarium 153-EDTMP or SM153-EDTMP (Quadramet) is a beta-particle-emitting, bone-seeking radiopharmaceutical. It is injected into a vein and distributed throughout the body, where it is absorbed in areas where cancer has invaded the bone.

Clinical trials

A clinical trial is a type of medical research study. After being developed and tested in a laboratory, potential new ways of fighting cancer need to be studied in people. If found to be safe and effective in a clinical trial, a drug, device, or treatment approach may be approved by the U.S. 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. Treatment trials are done in phases.

- **Phase I trials** study the dose, safety, and side effects of an investigational drug or treatment approach. They also look for early signs that the drug or approach is helpful.
- **Phase II trials** study how well the drug or approach works against a specific type of cancer.
- **Phase III trials** test the drug or approach against a standard treatment. If the results are good, it may be approved by the FDA.
- **Phase IV trials** study the long-term safety and benefit of an FDA-approved treatment.

Who can enroll?

Every clinical trial has rules for joining, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, or general health. These requirements 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 group of experts called a research team. The research team 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 with family, friends, or others whom you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

Start the conversation

Don't wait for your doctor to bring up clinical trials. Start the conversation and learn about all of your treatment options. If you find a study that you may be eligible for, ask your treatment team if you meet the requirements. If you have already started standard treatment you may not be eligible for certain clinical trials. Try not to be discouraged if you cannot join. New clinical trials are always becoming available.

Frequently asked questions

There are many myths and misconceptions surrounding clinical trials. The possible benefits and risks are not well understood by many with cancer.

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. You may, however, have costs indirectly related to the trial, such as the cost of transportation or child care due to extra appointments. During the trial, you will continue to receive standard cancer care. This care is billed to—and often covered by—insurance. You are responsible for copays and any costs for this care that are not covered by your insurance.



Finding a clinical trial

In the United States

NCCN Cancer Centers

[NCCN.org/cancercenters](https://www.nccn.org/cancercenters)

The National Cancer Institute (NCI)

[cancer.gov/about-cancer/treatment/clinical-trials/search](https://www.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](https://www.cancer.gov/contact)

Other treatments

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, which 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. A catheter will be inserted into an artery and guided to the tumor. Once in place, beads will be inserted to block the blood flow. With chemoembolization, the beads are coated with chemotherapy. Radioembolization uses small radioactive beads.

Supportive care

Supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life. It might include pain relief (palliative care), emotional or spiritual support, financial aid, or family counseling. Supportive care is given during all cancer stages. Tell your care team how you are feeling and about any side effects. Best supportive care is used with other treatments to improve quality of life. Best supportive care, supportive care, and palliative care are often used interchangeably.

It is important to tell your care team about all side effects so they can be managed.

Key points

- Treatment decisions should involve a multidisciplinary team (MDT) or a team of health care professionals from different fields of medicine who have knowledge (expertise) and experience with your type of cancer.
- Surgery to remove a metastasis is called a metastasectomy. This is different from a mastectomy, which is surgery to remove the breast.
- 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.



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](https://www.nccn.org/patients/response)

5

Chondrosarcoma

- 39 Overview
- 39 Treatment
- 40 Recurrence
- 40 Metastatic disease
- 41 Key points

Chondrosarcoma starts in the cells that form cartilage. Treatment is usually surgery to remove the tumor. Radiation therapy might be done in some cases. 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 grade of the tumor and if it can be removed with surgery.

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.

Treatment is grouped as follows:

- Low grade and intracompartmental (intracompartmental tumor has not grown outside of the bone)
- High grade (grade 2 or 3), clear cell, or extracompartmental (tumor has grown outside of the bone)
- Metastatic disease at initial diagnosis
- Dedifferentiated – treated with Ewing sarcoma therapy
- Mesenchymal – treated with Ewing sarcoma therapy

Treatment

Treatment options will be based on your wishes and your care team's recommendations.

Low-grade and intracompartmental chondrosarcoma

A low-grade (grade 1) and intracompartmental tumor is still within the bone. It has not grown through the bone wall or spread to any other part of the body. These tumors are sometimes called atypical cartilaginous tumors.

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.

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

- Physical exam
- X-rays of primary site and/or MRI or CT (both with contrast) as needed every 6 to 12 months for 2 years, then yearly as needed
- Chest imaging every 6 to 12 months for 2 years, then yearly as needed

High-grade, clear cell, or extracompartmental chondrosarcoma

A tumor that is high grade (grade 2 or 3) looks very abnormal under a microscope. Clear cell is a type of high-grade chondrosarcoma. An extracompartmental bone cancer has grown through the bone wall and out of the area where it started.

Treatment options include:

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

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

- Physical exam
- X-rays of primary site and/or MRI or CT (both with contrast) 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 recurrence.

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 if there are a few metastases called oligometastases or if the cancer is widespread. 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 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

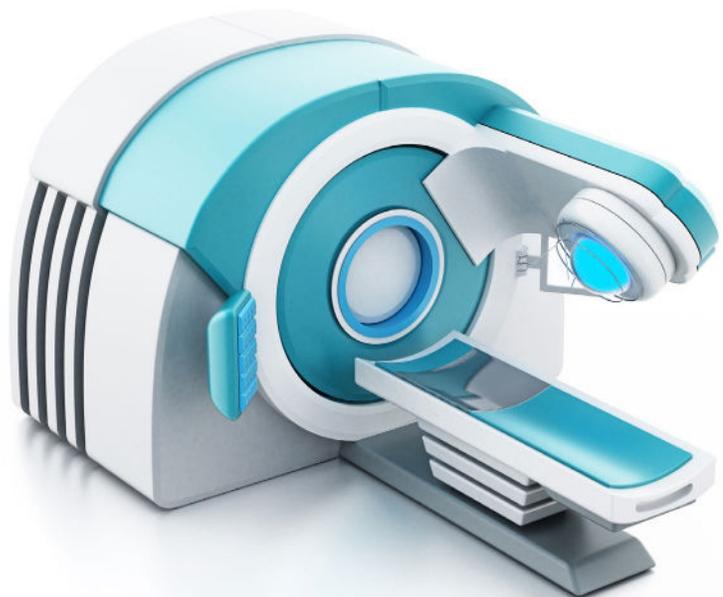
Before starting systemic therapy, tumor mutational burden (TMB) and biomarker testing might be done. Not everyone with metastatic chondrosarcoma is able to have surgery or systemic therapy. Ivosidenib (Tibsovo) would be an option for those who have *IDH1* mutations.

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 interlesional excision or curettage. Surgery to remove all of a tumor is called wide excision.
- 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.

Radiation therapy

Radiation therapy uses high-energy radiation from x-rays, gamma rays, protons, and other sources to kill cancer cells and shrink tumors. It is also used to treat pain caused by cancer.



6

Chordoma

- 43 Overview
- 43 Treatment
- 44 Monitoring
- 44 Recurrence
- 46 Key points

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.

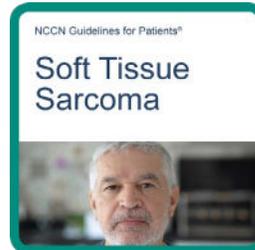
Overview

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 information on soft tissue sarcomas at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](https://www.nccn.org/patientguidelines) app.



Treatment

Chordomas are treated based on the location of the tumor:

- Tumors in the spine that include the sacrum and coccyx
- 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:

- If a tumor is resectable, RT might be given before, during, or after surgery.
- If a tumor is unresectable, then RT might be an option.

Tumors in skull base

For tumors at or in the base of the skull:

- If a tumor is resectable, 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 a 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 a physical exam and imaging tests for up to 10 years or as needed.

Get to know your care team and help them get to know you.

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 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 surgery, systemic therapy, or radiation therapy. These treatments might be used alone or in combination. Systemic therapy is drug therapy that works throughout the body. Before starting systemic therapy, tumor mutational burden (TMB) and biomarker testing might be done. Most systemic therapy options for chordoma recurrence are targeted therapies. Best supportive care is given with metastatic recurrence. Best supportive care is treatment to improve quality of life.

For recurrence treatment options, **see Guide 3 and 4.**

Guide 3

Treatment options: Chordoma recurrence

Local recurrence

- Surgical excision and/or
- Radiation therapy and/or
- Systemic therapy (see Guide 4)

Metastatic recurrence

- Systemic therapy (see Guide 4) and/or
- Surgical excision and/or
- Radiation therapy and/or
- Best supportive care

Guide 4

Systemic therapy options: Recurrence

Preferred

—

Other recommended

- Imatinib
- Dasatinib
- Sunitinib

In some cases

- Imatinib with cisplatin or sirolimus
- Erlotinib
- Lapatinib if EGFR-positive
- Sorafenib

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 treatment to improve quality of life.

Keep a pain diary

A pain diary is a written record that helps you keep track of when you have pain, how bad it is, what causes it, and what makes it better or worse. Use a pain diary to discuss your pain with your care team. You might be referred to a specialist for pain management.

Include in your pain diary:

- ✓ The time and dose of all medicines
- ✓ When pain starts and ends or lessens
- ✓ Where you feel pain
- ✓ Describe your pain. Is it throbbing, sharp, tingling, shooting, or burning? Is it constant, or does it come and go?
- ✓ Does the pain change at different times of day? When?
- ✓ Does the pain get worse before or after meals? Does certain food or drink make it better?
- ✓ Does the pain get better or worse with activity? What kind of activity?
- ✓ Does the pain keep you from falling asleep at night? Does pain wake you up in the night?
- ✓ Rate your pain from 0 (no pain) to 10 (worst pain you have ever felt)
- ✓ Does pain get in the way of you doing the things you enjoy?

7

Ewing sarcoma

- 48 Overview
- 48 Testing
- 49 Primary treatment
- 50 Local control treatment
- 50 Disease progression
- 51 Metastatic treatment
- 52 Key points

Ewing sarcoma is a group of cancers that start in the bone or soft tissue. Treatment begins with chemotherapy. 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, to other bones, or to the bone marrow.

Testing

Before starting treatment, you will have imaging, blood, 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 5**.

Guide 5

Testing for Ewing sarcoma

Medical history and physical exam

MRI with or without CT (both with contrast) of primary site

Chest CT

PET/CT (head-to-toe) 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)

Fertility consultation should be considered

Primary treatment

Chemotherapy is the first or primary treatment for all types of Ewing sarcoma. It will include a combination of chemotherapies. This is called multiagent chemotherapy. You will have at least 9 weeks of multiagent first-line chemotherapy. **See Guide 6.**

Restaging

After primary treatment of first-line chemotherapy, 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
- 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 6

First-line systemic therapy options

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

Local control treatment

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

Local control therapy might be:

- Surgery to remove the tumor (wide excision). Limb-sparing surgery or amputation might be possible.
- Radiation therapy (RT)
- Combination RT and surgery
- Chemotherapy after local control treatment

With local control treatment, the goal is to surgically remove all cancer or destroy all cancer cells with RT. Sometimes, both surgery and RT are needed. Chemotherapy might be given to complete the full course of treatment. The best treatment for each person with Ewing sarcoma is determined by a multidisciplinary cancer team and shared decision-making with the person with cancer and/or family.

Monitoring

After completing local control treatment, you will be monitored to watch 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 chemotherapy with the possibility of more surgery or radiation therapy.

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, then options include a second-line chemotherapy or best supportive care. Supportive care aims to improve quality of life.

For second-line systemic therapy options, **see Guide 7**.

Guide 7

Second-line systemic therapy options: Recurrence or disease progression

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.

Used in some cases

- Ifosfamide, carboplatin, and etoposide. Vincristine might be added.
- Lurbinectedin

Metastatic treatment

Metastatic Ewing sarcoma is cancer found throughout the body. Treatment for an initial diagnosis of metastatic Ewing sarcoma is different than cancer that has spread during or after treatment known as progressive or recurrent disease. This section is for those with metastatic Ewing sarcoma at diagnosis.

Treatment focuses on the primary (main) site and/or the distant metastases. In other words, treatment might include surgery or radiation therapy (RT) of the main site with treatment for the metastatic sites. Systemic therapy is possible. Before starting systemic therapy, tumor mutational burden (TMB) and biomarker testing might be done.

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 a lung metastasis might include surgery and/or RT.

For treatment of an initial diagnosis of metastatic Ewing sarcoma, **see Guide 8**.

Local control

Treatment that focuses on the primary (main) tumor is called local control therapy as described earlier.

Widely metastatic

Treatment for widely metastatic disease might include chemotherapy, palliative surgery, or palliative radiation. Other techniques might be used to treat metastases.

Guide 8

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

- Chemotherapy is the first or primary treatment for all types of Ewing sarcoma.
- Treatment that focuses on the main tumor site(s) in Ewing sarcoma is called local control therapy. It follows chemotherapy. The goal is to remove all cancer or destroy all cancer cells.
- Metastatic cancer is cancer found throughout the body. Treatment for an initial diagnosis of metastatic Ewing sarcoma is different than cancer that has spread during or after treatment known as progressive or recurrent disease.
- Treatment for widely metastatic disease might include chemotherapy, palliative surgery, or palliative radiation.

Did you know?

The terms “chemotherapy” and “systemic therapy” are often used interchangeably, but they are not the same. Chemotherapy, targeted therapy, and immunotherapy are all types of systemic therapy.

"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."



8

Giant cell tumor of bone

- 54 Overview
- 55 Localized disease
- 55 Metastatic disease
- 56 Monitoring
- 56 Recurrence
- 56 Key points

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.

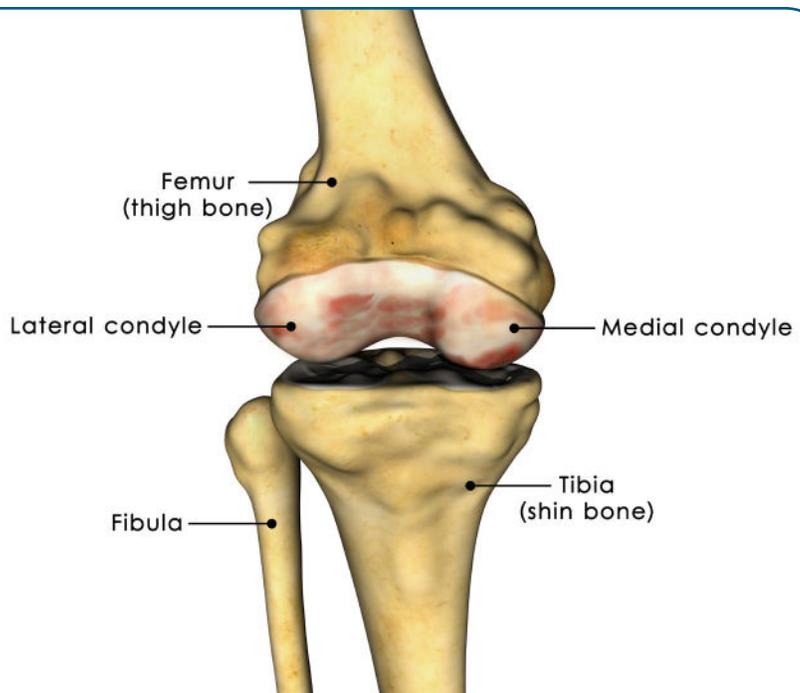
It is not known what causes a giant cell tumor. GCTB usually develops near a joint at the end of the bone. The knee is the most common location, but it can be found in the bones of the arms and the legs. It can also affect the flat bones, such as the breastbone or pelvis.

GCTB has a strong tendency to return to the same location. This is called local recurrence. 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 cancer is in one area (localized) or has spread to distant sites (metastasized).

Giant cell tumor of bone

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



Localized disease

In localized disease, cancer is confined to one area. Due to the location of the tumor or if you have other more serious health issues, surgery might not be possible. If this is the case, see unresectable treatment options.

Resectable

A resectable tumor can be removed with surgery. When possible, you will have surgery to remove the tumor. Frequently, first-line 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. It might include those with a resectable tumor, but who have other more serious health conditions that prevent surgery.

Treatments include:

- Denosumab (preferred) and/or
- Embolization (preferred) and/or
- Radiation therapy (RT)

There are some risks with RT and long-term use of denosumab. You should see a dentist before starting denosumab. Discuss with your care team which treatment is right for you.

After treatment you will have imaging tests, such as x-rays, CT, and MRI. If tests find that

Your preferences about treatment are always important. Talk to your care team and make your wishes known.

the tumor is now resectable, you will have surgery to remove the tumor.

Metastatic disease

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

For resectable tumors, you might have surgery to remove the main tumor and metastasis.

For unresectable tumors, options include:

- Denosumab
- Radiation therapy
- Observation

You should see a dentist before starting denosumab. Observation is sometimes referred to as watch-and-wait. Ask what this might mean for you.

Monitoring

After completing treatment, you will be monitored to watch for signs that cancer has returned called recurrence. Monitoring includes physical exam and 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. Treatment is based on if it is a local or metastatic recurrence. For metastatic recurrence, see *Metastatic disease* in this chapter.

Local recurrence

For a resectable tumor, you might have chest imaging to see if there are any metastases in the lungs. Denosumab might be given before surgery to remove the primary tumor (wide excision). Both resectable and unresectable treatment will follow *Localized disease* in this chapter.

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 that cancer 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.

9

Osteosarcoma

- 58 Overview
- 58 Testing
- 59 Systemic therapy
- 60 High-grade osteosarcoma
- 60 Metastatic disease at diagnosis
- 61 Periosteal and parosteal osteosarcoma
- 61 Monitoring
- 61 Relapse
- 62 Key points

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. In some cases, osteosarcoma runs in families. 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 types of osteosarcoma:

- Intramedullary (inside the bone)
- Surface (on the bone surface)
- Extraskelatal (outside the bone known as soft tissue sarcoma)

For treating extraskelatal osteosarcomas, see information on soft tissue sarcomas at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.

Testing

Before starting treatment, you will have imaging and blood 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 9**.

Guide 9 Testing for osteosarcoma

Medical history and physical exam

MRI with or without CT (both with contrast) of primary site

Chest imaging including chest CT

PET/CT (head-to-toe) and/or bone scan

MRI or CT (both with contrast) of skeletal and other metastatic sites

Lactate dehydrogenase (LDH)

Alkaline phosphatase (ALP)

Fertility consultation should be considered

Consider personal and family history for genetic consultation and testing

Systemic therapy

Chemotherapy and other systemic therapies are often part of treatment for osteosarcoma. First-line chemotherapies are those tried first. Second-line systemic therapy options are used when disease progresses or does not respond

to first-line options. **See Guide 10** and **Guide 11**.

Guide 10

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 11

Second-line systemic therapy options

Preferred options

- Ifosfamide (high dose) with or without etoposide
- Regorafenib
- Sorafenib

Other recommended

- Cabozantinib
- Cyclophosphamide and topotecan
- Docetaxel and gemcitabine
- Gemcitabine
- Sorafenib with everolimus

Used in some cases

- Cyclophosphamide and etoposide
- Ifosfamide, carboplatin, and etoposide
- High-dose methotrexate
- High-dose methotrexate, etoposide, and ifosfamide
- Sm153-EDTMP for relapsed or refractory disease beyond second-line therapy

High-grade osteosarcoma

This section is for 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. Treatment involves chemotherapy, then surgery, followed by more chemotherapy.

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 an imaging test to restage your cancer, assess treatment response, and plan surgery.

Surgery or local control therapy

Most osteosarcomas are treated with surgery to remove all of the cancer cells in your body. MRI and CT scans help determine if the osteosarcoma can be removed with surgery. Most osteosarcomas are resectable with limb-sparing surgery. 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 ablation.

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 chemotherapy. When cancer remains in the surgical margin, it is called a positive margin. Treatment might be chemotherapy, 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, tumor mutational burden (TMB) and biomarker testing might be done.

Periosteal and parosteal osteosarcoma

Periosteal osteosarcoma

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

Low-grade osteosarcoma or parosteal osteosarcoma

Treatment for low-grade osteosarcoma is surgery to remove the tumor called wide excision. Chemotherapy 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.

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 orthopedist 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 11**.

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 (resection), if possible
- Clinical trial
- Palliative radiation therapy (may include SM153-EDTMP)
- Best supportive care

Best supportive care is treatment to improve quality of life.

Key points

- Osteosarcoma is the most common type of bone cancer. It usually occurs around the knee or shoulder.
- Treatment most commonly involves chemotherapy 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.
- 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.



Take our [survey](#)
and help make the
NCCN Guidelines for Patients
better for everyone!

[NCCN.org/patients/comments](https://www.nccn.org/patients/comments)

10

Making treatment decisions

- 64 It's your choice
- 64 Questions to ask
- 74 Resources

It's important to be comfortable with the cancer treatment you choose. This choice starts with having an open and honest conversation with your care team.

It's your choice

In shared decision-making, you and your doctors share information, discuss the options, and agree on a treatment plan. It starts with an open and honest conversation between you and your care team.

Treatment decisions are very personal. What is important to you may not be important to someone else.

Some things that may play a role in your decision-making:

- What you want and how that might differ from what others want
- Your religious and spiritual beliefs
- Your feelings about certain treatments
- Your feelings about pain or side effects
- Cost of treatment, travel to treatment centers, and time away from school or work
- Quality of life and length of life
- How active you are and the activities that are important to you

Think about what you want from treatment. Discuss openly the risks and benefits of specific treatments and procedures. Weigh options and share concerns with your care team. If you take the time to build a relationship with your

care team, it will help you feel supported when considering options and making treatment decisions.

Second opinion

It is normal to want to start treatment as soon as possible. While cancer can't be ignored, there is time to have another doctor review your test results and suggest a treatment plan. This is called getting a second opinion, and it's a normal part of cancer care. Even doctors get second opinions!

Things you can do to prepare:

- Check with your insurance company about its rules on second opinions. There may be out-of-pocket costs to see doctors who are not part of your insurance plan.
- Make plans to have copies of all your records sent to the doctor you will see for your second opinion.

Support groups

Many people diagnosed with cancer find support groups to be helpful. Support groups often include people at different stages of treatment. Some people may be newly diagnosed, while others may be finished with treatment. If your hospital or community doesn't have support groups for people with cancer, check out the websites listed in this book.

Questions to ask

Possible questions to ask your care team are listed on the following pages. Feel free to use some of these questions or come up with your own. Be clear about your goals for treatment and find out what to expect from treatment.

Resources

American Association for Cancer Research (AACR)
aacr.org

American Cancer Society (ACS)
cancer.org/cancer

American Society of Clinical Oncology (ASCO)
cancer.net/cancer-types/bone-cancer-sarcoma-bone

Be The Match®
bethematch.org

Blood & Marrow Transplant Information Network (BMT InfoNet)
bmtinfonet.org

CancerCare
cancercare.org

Cancer Hope Network
cancerhopenetwork.org

Cancer Support Community
cancersupportcommunity.org/living-cancer

Chemocare
chemocare.com

Children's National®
childrensnational.org

Children's Oncology Group
survivorshipguidelines.org

Chordoma Foundation
chordomafoundation.org

KidsHealth® (also available en español)
kidshealth.org

MedlinePlus (also available en español)
medlineplus.gov

Musculoskeletal Tumor Society (MSTS)
msts.org

My Survival Story
mysurvivalstory.org

National Bone Marrow Transplant Link
nbmtlink.org

National Cancer Institute (NCI)
cancer.gov/types

National Coalition for Cancer Survivorship
canceradvocacy.org/resources/cancer-survival-toolbox

National Financial Resource Directory - Patient Advocate Foundation
patientadvocate.org/explore-our-resources/national-financial-resource-directory

National Hospice and Palliative Care Organization
nhpco.org/patients-and-caregivers

National Leiomyosarcoma Foundation
nlmsf.org

National Organization for Rare Disorders (NORD)
rarediseases.org

Northwest Sarcoma Foundation
nwsarcoma.org

OncoLink (also available en español)
oncolink.org

The Osteosarcoma Institute
osinst.org

Patient Access Network Foundation
panfoundation.org

Radiological Society of North America
(RSNA)
radiologyinfo.org

SARC
sarctrials.org

Sarcoma Foundation of America (SFA)
curesarcoma.org

Stupid Cancer
stupidcancer.org

Testing.com
testing.com

The Alan B. Slifka Foundation
slifkafoundation.org

The Paula Takacs Foundation
paulatakacsfoundation.org

The QuadW Foundation
quadw.org



Let us know what you think!

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

NCCN.org/patients/response



Words to know

ablation

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

biomarker testing

A 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.

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 parent to child through coded information in cells (genes).

hypercalcemia

Higher than normal levels of calcium in the blood.

histology

The study of tissues and cells under a microscope.

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.

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 sometimes 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.

stereotactic radiosurgery (SRS)

A cancer treatment that uses special equipment to position the body and give one precise, large dose of radiation.

subtype

A smaller group within a type of cancer that is based on certain cell features.

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.

NCCN Contributors

This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Bone Cancer, Version 2.2023. It was adapted, reviewed, and published with help from the following people:

Dorothy A. Shead, MS
Senior Director
Patient Information Operations

Tanya Fischer, MEd, MSLIS
Senior Medical Writer

Laura Phillips
Graphic Artist

The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Bone Cancer, Version 2.2023 were developed by the following NCCN Panel Members:

J. Sybil Biermann, MD/Chair
University of Michigan Rogel Cancer Center

Angela Hirbe, MD, PhD/Vice-Chair
Siteman Cancer Center at Barnes-Jewish Hospital and Washington University School of Medicine

Mark Agulnik, MD
City of Hope National Medical Center

Nicholas M. Bernthal, MD
UCLA Jonsson Comprehensive Cancer Center

Sarah Boles, MD
UC San Diego Moores Cancer Center

Brian Brigman, MD, PhD
Duke Cancer Institute

***Alexandra K. Callan, MD**
UT Southwestern Simmons Comprehensive Cancer Center

Lee D. Cranmer, MD, PhD
Fred Hutchinson Cancer Center

Saeed Dianat, MD
Abramson Cancer Center at the University of Pennsylvania

Eric Donnelly, MD
Robert H. Lurie Comprehensive Cancer Center of Northwestern University

Nicola Fabbri, MD
Memorial Sloan Kettering Cancer Center

***Michael Ferguson, MD, MS**
Indiana University Melvin and Bren Simon Comprehensive Cancer Center

Patrick Getty, MD
Case Comprehensive Cancer Center/ University Hospitals Seidman Cancer Center and Cleveland Clinic Taussig Cancer Institute

John Groundland, MD
Huntsman Cancer Institute at the University of Utah

Susan M. Hiniker, MD
Stanford Cancer Institute

Margo L. Hoover-Regan, MD
University of Wisconsin Carbone Cancer Center

Lisa Kafchinski, MD
O'Neal Comprehensive Cancer Center at UAB

Joseph B. Kuechle, MD, PhD
Roswell Park Comprehensive Cancer Center

Sameera Kumar, MD
Fox Chase Cancer Center

Dieter Lindskog, MD
Yale Cancer Center/Smilow Cancer Hospital

David R. Lucas, MD
University of Michigan Rogel Cancer Center

***Joel L. Mayerson, MD**
The Ohio State University Comprehensive Cancer Center - James Cancer Hospital and Solove Research Institute

Sean V. McGarry, MD
Fred & Pamela Buffett Cancer Center

Carol D. Morris, MD, MS
The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins

Damon R. Reed, MD
Moffitt Cancer Center

Peter S. Rose, MD
Mayo Clinic Cancer Center

Victor M. Santana, MD
St. Jude Children's Research Hospital/The University of Tennessee Health Science Center

Robert L. Satcher, MD, PhD
The University of Texas MD Anderson Cancer Center

Joseph Schwab, MD, MS
Massachusetts General Hospital Cancer Center

Herbert Schwartz, MD
Vanderbilt-Ingram Cancer Center

Steven W. Thorpe, MD
UC Davis Comprehensive Cancer Center

Breelyn A. Wilky, MD
University of Colorado Cancer Center

Rosanna L. Wustrack, MD
UCSF Helen Diller Family Comprehensive Cancer Center

NCCN

Lisa E. Hang, PhD
Oncology Scientist/Medical Writer

Mary Anne Bergman
Guidelines Coordinator

* Reviewed this patient guide. For disclosures, visit [NCCN.org/disclosures](https://www.nccn.org/disclosures).

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
800.641.2422 • UH Seidman Cancer Center
uhhospitals.org/services/cancer-services
866.223.8100 • CC Taussig Cancer Institute
my.clevelandclinic.org/departments/cancer
216.844.8797 • Case CCC
case.edu/cancer

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

Dana-Farber/Brigham and Women's
Cancer Center | Massachusetts
General Hospital Cancer Center
Boston, Massachusetts
617.732.5500 • 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 • huntsmancancer.org

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

Mayo Clinic 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 Sidney Kimmel Comprehensive
Cancer Center at Johns Hopkins
Baltimore, Maryland
410.955.8964
www.hopkinskimmelcancercenter.org

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 • cancer.ucla.edu

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

Index

- ablation** 36
- amputation** 31
- appendicular skeleton** 23
- best supportive care** 36
- biomarker tests** 18–19
- biopsy** 17
- bone tumors** 7
- brachytherapy** 33
- cancer stages** 22–27
- chemotherapy** 32
- clinical trials** 34–35
- cryosurgery or cryotherapy** 36
- curettage** 39
- embolization** 36
- excision** 39
- external beam radiation therapy (EBRT)** 33
- family history** 12, 17, 19
- fertility** 11
- genetic tests** 17
- grade** 26
- immunotherapy** 33
- internal radiation** 33
- limb-sparing surgery** 30
- local control therapy** 50–51
- MSI-H/dMMR** 18
- multidisciplinary team (MDT)** 29–30
- mutation** 18–19
- orthopedic** 29–30
- pain** 46
- pelvis** 25
- performance status (PS)** 13
- pregnancy** 11
- radiation therapy (RT)** 36
- radioactive beads or seeds** 33, 36
- radiofrequency ablation (RFA)** 36
- remodeling** 6
- sarcoma** 7
- SM153-EDTMP** 33
- soft tissue sarcoma** 7
- spine** 24
- stereotactic radiosurgery (SRS)** 33
- supportive care** 36
- surgery** 30–31
- systemic therapy** 32–33
- targeted therapy** 32
- translocation** 18
- tumor mutational burden (TMB)** 19
- tumor tests** 18–19





NCCN
GUIDELINES
FOR PATIENTS®

Bone Cancer

2023

To support the NCCN Guidelines for Patients, visit [NCCN.Foundation.org/Donate](https://www.nccn.org/Donate)

[NCCNFoundation.org/Donate](https://www.nccn.org/Donate)



National Comprehensive
Cancer Network®

3025 Chemical Road, Suite 100
Plymouth Meeting, PA 19462
215.690.0300

[NCCN.org/patients – For Patients](https://www.nccn.org/patients) | [NCCN.org – For Clinicians](https://www.nccn.org)