Bone Cancer

What is cancer?

The body is made up of trillions of living cells. Normal body cells grow, divide into new cells, and die in an orderly fashion. During the early years of a person's life, normal cells divide faster to allow the person to grow. After the person becomes an adult, most cells divide only to replace worn-out or dying cells or to repair injuries.

Cancer begins when cells in a part of the body start to grow out of control. There are many kinds of cancer, but they all start because of out-of-control growth of abnormal cells.

Cancer cell growth is different from normal cell growth. Instead of dying, cancer cells continue to grow and form new, abnormal cells. Cancer cells can also invade (grow into) other tissues, something that normal cells cannot do. Growing out of control and invading other tissues are what makes a cell a cancer cell.

Cells become cancer cells because of damage to DNA. DNA is in every cell and directs all its actions. In a normal cell, when DNA gets damaged the cell either repairs the damage or the cell dies. In cancer cells, the damaged DNA is not repaired, but the cell doesn’t die like it should. Instead, this cell goes on making new cells that the body does not need. These new cells will all have the same damaged DNA as the first cell does.

People can inherit damaged DNA, but most DNA damage is caused by mistakes that happen while the normal cell is reproducing or by something in our environment. Sometimes the cause of the DNA damage is something obvious, like cigarette smoking. But often no clear cause is found.

In most cases the cancer cells form a tumor. Some cancers, like leukemia, rarely form tumors. Instead, these cancer cells involve the blood and blood-forming organs and circulate through other tissues where they grow.

Cancer cells often travel to other parts of the body, where they begin to grow and form new tumors that replace normal tissue. This process is called metastasis. It happens when the cancer cells get into the bloodstream or lymph vessels of our body.

No matter where a cancer may spread, it is always named for the place where it started. For example, breast cancer that has spread to the liver is still called breast cancer, not liver cancer. Likewise, prostate cancer that has spread to the bone is metastatic prostate cancer, not bone cancer.

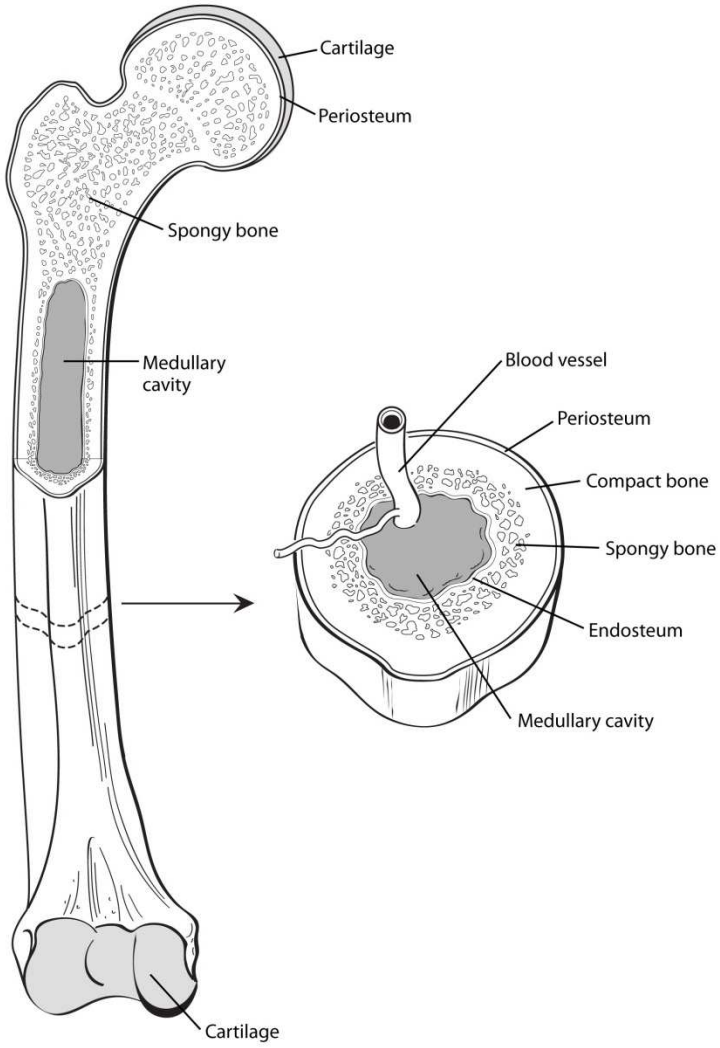
Different types of cancer can behave very differently. For example, lung cancer and breast cancer are very different diseases. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

Not all tumors are cancerous. Tumors that aren’t cancer are called benign. Benign tumors can cause problems – they can grow very large and press on healthy organs and tissues. But they cannot grow into (invade) other tissues. Because they can’t invade, they also can’t spread to other parts of the body (metastasize). These tumors are almost never life threatening.

What is bone cancer?

Normal bone tissue

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



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

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

Most bones start out as cartilage. The body then lays calcium down onto the cartilage to form bone. After the bone is formed, some cartilage may remain at the ends to act as a cushion between bones. This cartilage, along with ligaments and some other tissues connect bones to form a joint. In adults, cartilage is mainly found at the end of some bones as part of a joint. It

is also seen at the place in the chest where the ribs meet the sternum (breastbone) and in parts of the face. The trachea (windpipe), larynx (voice box), and the outer part of the ear are other structures that contain cartilage.

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

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

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

Types of bone tumors

Most of the time when someone with cancer is told they have cancer in the bones, the doctor is talking about a cancer that has spread to the bones from somewhere else. This is called *metastatic cancer.* It can be seen in many different types of advanced cancer, like breast cancer, prostate cancer, and lung cancer. When these cancers in the bone are looked at under a microscope, they look like the tissue they came from. For example, if someone has lung cancer that has spread to bone, the cells of the cancer in the bone still look and act like lung cancer cells. They do not look or act like bone cancer cells, even though they are in the bones. Since these cancer cells still act like lung cancer cells, they still need to be treated with drugs that are used for lung cancer. For more information about metastatic bone cancer, please see our document called *Bone Metastasis,* as well as the document on the specific place the cancer started (*Breast Cancer*, *Lung Cancer*, *Prostate Cancer,* etc.).

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

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

There are several different types of bone tumors. Their names are based on the area of bone or surrounding tissue that is affected and the kind of cells forming the tumor. Some primary

bone tumors are *benign* (not cancerous), and others are *malignant* (cancerous). Most bone cancers are sarcomas.

Benign bone tumors

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

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

These benign tumors are not discussed further in this document, which is limited to bone cancers.

Malignant bone tumors

Osteosarcoma: Osteosarcoma (also called *osteogenic sarcoma*) is the most common primary bone cancer. This cancer starts in the bone cells. It most often occurs in young people between the ages of 10 and 30, but about 10% of osteosarcoma cases develop in people in their 60s and 70s. It is rare in middle aged people, and is more common in males than females. These tumors develop most often in bones of the arms, legs, or pelvis. This cancer is not discussed in detail in this document, but is covered in our document called *Osteosarcoma.*

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

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

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

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

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

- *Dedifferentiated chondrosarcomas* start out as typical chondrosarcomas but then some parts of the tumor change into cells like those of a high grade sarcoma (such as high grade forms of malignant fibrous histiocytoma, osteosarcoma, or fibrosarcoma). This variant of chondrosarcoma tends to occur in older patients and is more aggressive than usual chondrosarcomas.

- *Clear cell chondrosarcoma* is a rare variant that grows slowly. It rarely spreads to other parts of the body unless it has already come back several times in the original location.

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

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

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

Fibrosarcoma: This is another type of cancer that develops more often in "soft tissues" than it does from bones. Fibrosarcoma usually occurs in elderly and middle-aged adults. Leg, arm, and jaw bones are the ones most often affected.

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

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

Other cancers that develop in bones

Non-Hodgkin lymphomas

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

Multiple myelomas

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

What are the key statistics about bone cancer?

The American Cancer Society's estimates for cancer of the bones and joints for 2013 are:

- About 3,010 new cases will be diagnosed

- About 1,440 deaths from these cancers are expected.

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

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

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

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

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

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

What are the risk factors for bone cancer?

A *risk factor* is anything that affects your chance of getting a disease such as cancer. Different cancers have different risk factors. For example, exposing skin to strong sunlight is a risk factor for skin cancer. Smoking is a risk factor for cancers of the lung, mouth, larynx, bladder, kidney, and several other organs. But having a risk factor, or even several, does not mean that you will get the disease. Most people with bone cancers do not have any apparent risk factors.

Genetic disorders

A very small number of bone cancers (especially osteosarcomas) appear to be hereditary and are caused by defects (mutations) in certain genes.

Osteosarcomas

Children with certain rare inherited syndromes have an increased risk of developing osteosarcoma.

- The **Li-Fraumeni syndrome** makes people much more likely to develop several types of cancer, including breast cancer, brain cancer, osteosarcoma, and other types of sarcoma. Most of those cases are caused by a mutation of the *p53* tumor suppressor gene, but some are caused by mutations in the gene *CHEK2*.

- Another syndrome that includes bone cancer is the **Rothmund-Thomson syndrome**. Children with this syndrome are short, have skeletal problems, and rashes. They also are more likely to develop osteosarcoma. This syndrome is caused by abnormal changes in the gene *REQL4*.

- **Retinoblastoma** is a rare eye cancer of children that can be hereditary. The inherited form of retinoblastoma is caused by a mutation (abnormal copy) of the *RB1* gene. Those with this mutation also have an increased risk of developing bone or soft tissue sarcomas. Also, if radiation therapy is used to treat the retinoblastoma, the risk of osteosarcoma in the bones around the eye is even higher.

Finally, there are families with several members who have developed osteosarcoma without inherited changes in any of the known genes. The gene defects that may cause cancers in these families haven't been discovered yet.

Chondrosarcomas

Multiple exostoses (sometimes called *multiple osteochondromas*) syndrome is an inherited condition that causes many bumps on a person's bones. These bumps are made mostly of cartilage. They can be painful and cause bones to deform and/or fracture. This disorder is caused by a mutation in any one of the 3 genes *EXT1*, *EXT2*, or *EXT3*. Patients with this condition have an increased risk of chondrosarcoma.

An enchondroma is a benign cartilage tumor that grows into the bone. People who get many of these tumors have a condition called *multiple enchondromatosis*. They have an increased risk of developing chondrosarcomas.

Chordomas

Chordomas seem to run in some families. The genes responsible have not yet been found, but familial chordoma has been linked to changes on chromosome 7.

Patients with the inherited syndrome *tuberous sclerosis*, which can be caused by defects (mutations) in either of the genes *TSC1* and *TSC2*, seem to have a high risk of chordomas during childhood.

Paget disease

Paget disease is a benign (non-cancerous) but pre-cancerous condition that affects one or more bones. It results in formation of abnormal bone tissue and is mostly a disease of people older than 50. Affected bones are heavy, thick, and brittle. They are weaker than normal bones and more likely to fracture (break). Most of the time Paget disease is not life threatening. Bone cancer (usually osteosarcoma) develops in about 1% of those with Paget disease, usually when many bones are affected.

Radiation

Bone exposure to ionizing radiation may also increase the risk of developing bone cancer. A typical x-ray of a bone is not dangerous, but exposure to large doses of radiation does pose a risk. For example, radiation therapy to treat cancer can cause a new cancer to develop in one of the bones in the treatment area. Being treated at a younger age and/or being treated with higher doses of radiation (usually over 60 Gy) increases the risk of developing bone cancer.

Exposure to radioactive materials such as radium and strontium can also cause bone cancer because these minerals build up in bones.

Non-ionizing radiation, like microwaves, electromagnetic fields from power lines, cellular phones, and household appliances, does not increase bone cancer risk.

Bone marrow transplantation

Osteosarcoma has been reported in a few patients who have undergone bone marrow (stem cell) transplantation.

Injuries

People have wondered whether injury to a bone can cause cancer, but this has never been proven. Many people with bone cancer remember having hurt that part of their bone. Most doctors believe that this did not cause the cancer, but rather that the cancer caused them to remember the incident or that the injury drew their attention to that bone and caused them to notice a problem that had already been present for some time.

Do we know what causes bone cancer?

The exact cause of most bone cancers is not known. However, scientists have found that bone cancers are associated with a number of other conditions, which are described in the section on risk factors. Still, most people with bone cancers do not have any known risk factors. Research is underway to learn more about the causes of these cancers.

Scientists have made great progress in understanding how certain changes in a person's DNA can cause normal cells to become cancerous. DNA carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. However, DNA affects more than our outward appearance. It may influence our risks for developing certain diseases, including some kinds of cancer.

DNA is divided into units called *genes*. Genes carry the recipes for making proteins, the molecules that determine all cell functions. Some genes contain instructions to control when our cells grow and divide. Genes that promote cell division are called *oncogenes*. Others that slow down cell division or cause cells to die at the right time are called *tumor suppressor*

genes. Cancers can be caused by DNA mutations (defects) that activate oncogenes or inactivate tumor suppressor genes. Some people with cancer have DNA mutations that they inherited from a parent. These mutations increase their risk for the disease.

The DNA mutations that cause some inherited forms of bone cancers are known (see the section, "What are the risk factors for bone cancer?"). In many cases, genetic testing can be used to see if someone has one of these mutations.

Most bone cancers are not caused by inherited DNA mutations. They are the result of mutations acquired during the person's lifetime. These mutations may result from exposure to radiation or cancer-causing chemicals, but most often they occur for no apparent reason. These mutations are present only in the cancer cells and so cannot be passed on to the patient's children.

Scientists are making progress in understanding this process, but there are still some points that are not completely understood. As their knowledge increases, they hope to develop ways to better prevent and treat bone cancers.

Can bone cancer be prevented?

Changes in lifestyle can help prevent many types of cancer. At present, however, no known lifestyle changes can prevent bone cancers.

Can bone cancer be found early?

Tests are routinely used to detect early stages of some types of cancer (such as breast, cervical, colorectal, and skin cancer) before they cause symptoms. At this time, no special tests are routinely recommended to detect bone cancers early. The best strategy for early diagnosis is prompt attention to the signs and symptoms of this disease.

How is bone cancer diagnosed?

A patient's symptoms, physical exam, and results of imaging tests, and blood tests may suggest that bone cancer is present. But in most cases, doctors must confirm this suspicion by examining a tissue or cell sample under a microscope (known as a *biopsy*).

Other diseases, such as bone infections, can cause symptoms and imaging results that could be confused with bone cancer. Accurate diagnosis of a bone tumor often depends on combining information about its location (what bone is affected and even which part of the bone is involved), appearance on x-rays, and appearance under a microscope.

Since a single bone metastasis can have the same sign and symptoms as a primary bone tumor, many doctors require a biopsy to diagnose a patient's first bone metastasis. After that, additional bone metastases can usually be diagnosed based on x-rays and other imaging tests.

Signs and symptoms of bone cancer

Pain

Pain in the affected bone is the most common complaint of patients with bone cancer. At first, the pain is not constant. It may be worse at night or when the bone is used (for example, leg pain when walking). As the cancer grows, the pain will be there all the time. The pain increases with activity and the person might limp if a leg is involved.

Swelling

Swelling in the area of the pain may not occur until weeks later. It may be possible to feel a lump or mass depending on the location of the tumor.

Fractures

Bone cancer may weaken the bone it develops in, but most of the time the bones do not fracture (break). People with a fracture next to or through a bone cancer usually describe sudden severe pain in a limb that had been sore for a few months.

Other symptoms

Cancer can cause problems like weight loss and fatigue. If the cancer spreads to internal organs it may cause symptoms, too. For example, if the cancer spreads to the lung, you may have trouble breathing.

Bone pain and/or swelling are more often due to conditions other than cancer, such as injuries or arthritis. Still, if these problems go on for a long time without a known reason, you should see your doctor.

Imaging tests to detect bone cancer

X-rays

Most bone cancers show up on x-rays of the bone. The bone at the site of the cancer may appear "ragged" instead of solid. The cancer can also appear as a hole in the bone. Sometimes doctors can see a tumor around the defect in the bone that might extend into nearby tissues (such as muscle or fat). The radiologist (doctor who specializes in reading x-rays) can often tell if a tumor is malignant by the way it appears on the x-ray, but only a biopsy can absolutely determine that.

A chest x-ray is often done to see if bone cancer has spread to the lungs.

Computed tomography (CT) scans

The CT scan is an x-ray procedure that produces detailed, cross-sectional images of your body. Instead of taking one picture, like a conventional x-ray, a CT scanner takes many pictures as it rotates around you. A computer then combines these pictures into an image of a slice of your body. The machine creates multiple images of the part of your body that is being studied.

A CT scanner has been described as a large donut, with a narrow table in the middle opening. You will need to lie still on the table while the scan is being done. CT scans take longer than regular x-rays, and you might feel a bit confined by the ring while the pictures are being taken.

CT scans are helpful in staging cancer. They help tell if your bone cancer has spread into your lungs, liver, or other organs. These scans also show the lymph nodes and distant organs where metastatic cancer might be present.

Before the first set of pictures is taken you may be asked to drink 1 or 2 pints of a contrast agent. This helps outline the intestine so that certain areas are not mistaken for tumors. You may also receive an IV (intravenous) line through which a different kind of contrast dye is injected. This helps better outline structures in your body.

The injection can cause some flushing (redness and warm feeling that may last hours to days). A few people are allergic to the dye and get hives. Rarely, more serious reactions like trouble breathing and low blood pressure can occur. Medication can be given to prevent and treat allergic reactions. Be sure to tell the doctor if you have ever had a reaction to any contrast material used for x-rays or if you have an allergy to shellfish.

CT scans can also be used to precisely guide a biopsy needle into a suspected metastasis. For this procedure, called a CT-guided needle biopsy, the patient remains on the CT scanning table while a radiologist advances a biopsy needle toward the location of the mass. CT scans are repeated until the doctors are confident that the needle is within the mass. (See the section, "Needle biopsy.")

Magnetic resonance imaging (MRI) scans

MRI scans use radio waves and strong magnets instead of x-rays. The energy from the radio waves is absorbed and then released in a pattern formed by the type of tissue and by certain diseases. A computer translates the pattern of radio waves given off by the tissues into a very detailed image of parts of the body. Sometimes a contrast material called *gadolinium* is injected into your veins to better see the tumor.

MRI scans are often the best test for outlining a bone tumor. They are also particularly helpful for looking at the brain and spinal cord. MRI scans are a little more uncomfortable than CT scans. First, they take longer -- often up to an hour. Also, you have to be placed inside a tube, which is confining and can upset people with claustrophobia (fear of enclosed

spaces). The machine also makes a thumping noise that you may find disturbing. Some places provide headphones with music to block this out.

Radionuclide bone scans

This procedure helps show if a cancer has spread to other bones. It can find metastases earlier than regular x-rays. Bone scans also can show how much damage the primary cancer has caused in the bone.

For this test, the patient receives an injection of radioactive material called *technetium diphosphonate*. The amount of radioactivity used is very low and causes no long-term effects. This substance is attracted to diseased bone cells throughout the entire skeleton. Areas of diseased bone will be seen on the bone scan image as dense, gray to black areas, called "hot spots." These areas may suggest metastatic cancer is present, but arthritis, infection, or other bone diseases can also cause a similar pattern. To distinguish among these conditions, the cancer care team may use other imaging tests or take bone biopsies.

Positron emission tomography (PET or PET) scans

PET scans use glucose (a form of sugar) that contains a radioactive atom. A special camera can detect the radioactivity. Cancer cells absorb a lot of the radioactive sugar because of their high rate of metabolism. PET is useful to look for cancer throughout your body. PET scans can be more helpful than several different x-rays because it scans the whole body. It can sometimes help tell if a tumor is cancerous or benign. It is being combined with CT scans to better pinpoint some kinds of cancer.

Biopsy

A *biopsy* is a sample of tissue taken from a tumor so that it can be looked at under a microscope. This is the only way to know that the tumor is cancer and not some other bone disease. If cancer is present, the biopsy can tell the doctor if it is a primary bone cancer or cancer that started somewhere else and spread to the bone (metastasis). Several types of tissue and cell samples are used to diagnose bone cancer. It is very important a surgeon with experience in diagnosing and treating bone tumors do the biopsy procedure.

The surgeon will choose a biopsy method based on whether the tumor looks benign or malignant and exactly what type of tumor is most likely (based on the bone x-rays, the patient's age, and the location of the tumor). Some kinds of bone tumors can be recognized from needle biopsy samples, but larger samples (from a surgical biopsy) are often needed to diagnose other types. Whether the surgeon plans to remove the entire tumor at the time of the biopsy will also influence the choice of biopsy type. The wrong kind of biopsy can sometimes make it hard later for the surgeon to remove all of the cancer without having to also remove all or part of the arm or leg containing the tumor. It also may cause the cancer to spread.

Needle biopsy

There are 2 types of needle biopsies: *fine needle biopsies* and *core needle biopsies.* For both types, a local anesthetic is first used to numb the area for the biopsy. For fine needle aspiration (FNA), the doctor uses a very thin needle attached to a syringe to withdraw a small amount of fluid and some cells from the tumor mass. Sometimes, the doctor can aim the needle by feeling the suspicious tumor or area that is near the surface of the body. If the tumor cannot be felt because it is too deep, the doctor can guide the needle while viewing a CT scan. This is called a *CT guided needle biopsy* and it is often done by an x-ray specialist known as an *interventional radiologist.* In a core needle biopsy, the doctor uses a larger needle to remove a small cylinder of tissue (about 1/16 inch in diameter and 1/2 inch long). Many experts feel that a core needle biopsy is better than FNA to diagnose a primary bone cancer.

Surgical bone biopsy

In this procedure, a surgeon needs to cut through the skin to reach the tumor in order to remove a small piece of tissue. This is also called an *incisional biopsy*. If the entire tumor is removed (not just a small piece), it is called an *excisional biopsy*. These biopsies are often done under general anesthesia (with the patient asleep). They can also be done using a nerve block, which numbs a large area. If this type of biopsy is needed, it is important that the surgeon who will later remove the cancer be the one to do the biopsy.

How is bone cancer staged?

Staging is a process that tells the doctor how widespread a cancer may be. It will show whether the cancer has spread and how far. The treatment and prognosis (outlook) for bone cancers depend, to a large extent, on the patient's stage at diagnosis.

AJCC Staging System

One system that is used to stage all bone cancers is the American Joint Commission on Cancer (AJCC) system. It combines 4 factors to determine stage that go by the initials T, N, M, and G. **T** stands for features of **t**umor (its size and if it is in more than one spot on the bone), **N** stands for spread to lymph **n**odes, **M** is for **m**etastasis (spread) to distant organs, and **G** is for the tumor's **g**rade. The grade of a tumor is based on how abnormal the cells look when seen under a microscope. The higher the number, the more abnormal the cells appeared. Higher grade cancers tend to grow and spread more quickly than lower grade tumors.

This information about the tumor, lymph nodes, metastasis, and grade is combined in a process called *stage grouping*. The stage is then described in Roman numerals from I to IV (1-4).

T stages of bone cancer

TX: Primary tumor can't be measured

T0: No evidence of the tumor

T1: Tumor is 8 cm (around 3 inches) or less

T2: Tumor is larger than 8 cm

T3: Tumor is in more than one place on the same bone

N stages of bone cancer

N0: The cancer has not spread to the lymph nodes near the tumor

N1: The cancer has spread to nearby lymph nodes

M stages of bone cancer

M0: The cancer has not spread anywhere outside of the bone or nearby lymph nodes

M1: Distant metastasis (the cancer has spread)

- **M1a:** The cancer has spread only to the lung
- **M1b:** The cancer has spread to other sites (like the brain, the liver, etc.)

Grades of bone cancer

G1-G2: Low grade

G3-G4: High grade

TNM stage grouping

After the T, N, and M stages and the grade of the bone cancer have been determined, the information is combined and expressed as an overall stage. The process of assigning a stage number is called *stage grouping*. To determine the grouped stage of a cancer using the AJCC system, find the stage number below that contains the T, N, and M stages, and the proper grade.

Stage I: All stage I tumors are low grade and have not yet spread outside of the bone.

- **Stage IA:** T1, N0, M0, G1-G2: The tumor is 8 cm or less.
- **Stage IB:** T2 or T3, N0, M0, G1-G2: The tumor is either larger than 8 cm or it is in more than one place on the same bone.

Stage II: Stage II tumors have not spread outside the bone (like stage I) but are high grade.

- **Stage IIA:** T1, N0, M0, G3-G4: The tumor is 8 cm or less.
- **Stage IIB:** T2, N0, M0, G3-G4: The tumor is larger than 8 cm.

Stage III: T3, N0, M0, G3-G4: Stage III tumors have not spread outside the bone but are in more than one place on the same bone. They are high grade.

Stage IV: Stage IV tumors have spread outside of the bone they started in. They can be any grade.

- **Stage IVA:** Any T, N0, M1a, G1-G4: The tumor has spread to the lung.
- **Stage IVB:** Any T, N1, any M, G1-G4 OR Any T, any N, M1b, G1-G4: The tumor has spread to nearby lymph nodes or to distant sites other than the lung (or both).

Even though the AJCC staging system is widely accepted and used for most cancers, bone cancer specialists tend to simplify the stages into localized and metastatic. Localized includes stages I, II, and III, while metastatic is the same as stage IV.

Survival statistics for bone cancer

Survival rates are often used by doctors as a standard way of discussing a person's prognosis (outlook). Some patients with cancer may want to know the survival statistics for people in similar situations, while others may not find the numbers helpful, or may even not want to know them. If you do not want to read about the survival statistics for bone cancer given in the next few paragraphs, skip to the next section..

The 5-year survival rate refers to the percentage of patients who live *at least 5 years* after their cancer is diagnosed. Of course, many people live much longer than 5 years (and many are cured).

Five-year *relative* survival rates assume that some people will die of other causes and compare the observed survival with that expected for people without the cancer. This is a better way to see the impact of the cancer on survival.

In order to get 5-year survival rates, doctors have to look at people who were treated at least 5 years ago. Improvements in treatment since then may result in a more favorable outlook for people now being diagnosed with bone cancer.

Survival rates are often based on previous outcomes of large numbers of people who had the disease, but they cannot predict what will happen in any particular person's case. Many factors may affect a person's outlook, such as the type and grade of the cancer, the patient's age, where the cancer is located, the size of the tumor, and the treatment received. Your doctor can tell you how the numbers below may apply to you, as he or she is familiar with the aspects of your particular situation.

For all cases of bone cancer combined (in both adults and children), the 5-year relative survival is about 70%. For adults, the most common bone cancer is chondrosarcoma, which has a 5-year relative survival of about 80%. (Survival statistics for Ewing tumors and osteosarcoma can be found in our documents about those cancers.)

How is bone cancer treated?

This information represents the views of the doctors and nurses serving on the American Cancer Society's Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.

The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.

Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.

General treatment information

The main types of treatment for bone cancer are:

- Surgery
- Radiation
- Chemotherapy
- Targeted therapy

Often, more than one type of treatment is used. For information about some of the most common approaches used based on the extent of the disease, see the section “Treating specific bone cancers.”

Surgery for bone cancer

Surgery is the primary (main) treatment for most bone cancers. Surgery may also be needed to obtain a biopsy of the cancer. The biopsy and the surgical treatment are separate operations, but it is very important that they both be planned together. Ideally, the same surgeon should do both the biopsy and the main surgery. A biopsy taken from the wrong place can lead to problems when the surgeon does the operation to remove the cancer. Sometimes a poorly placed biopsy can even make it impossible to remove the cancer without cutting off the limb.

The main goal of surgery is to remove all of the cancer. If even a few cancer cells are left behind, they can grow and multiply to make a new tumor. To try to be sure that this doesn’t happen, surgeons remove the tumor plus some of the normal-appearing tissue that surrounds

it. This is known as *wide-excision*. Removing some normal-looking tissue helps ensure that all of the cancer is removed. After surgery, a pathologist will look at the tissue that was removed under the microscope to see if the margins (outer edges) contain cancer cells. If cancer cells were seen at the edges of the tissue, the margins are called *positive*. Positive margins can mean that some cancer was left behind. When no cancer is seen at the edges of the tissue, the margins are said to be *negative*, *clean*, or *clear*. A wide-excision with clean margins minimizes the risk that the cancer will grow back where it started.

Tumors in the arms or legs

Sometimes the entire limb needs to be removed in order to do a good wide-excision (and remove all of the cancer). This operation is known as an *amputation*. But most of the time the surgeon can remove the cancer without amputation. This is called *limb-salvage* or *limb-sparing surgery*. In going over treatment options, it is important to realize that there are advantages and disadvantages that go along with either type of surgery. For example, although for many people limb-salvage seems more acceptable than amputation, it is more complex and so can have more complications. Both operations have the same overall survival rates when done by expert surgeons. Studies looking at quality of life have shown little difference in how people react to the final result of the different procedures. Perhaps the biggest concern was seen in teenagers who worry about the social effects of their operation. Emotional issues can be very important and support and encouragement are needed for all patients.

No matter which type of surgery is done, rehabilitation will be needed afterward. This can be the hardest part of treatment. If possible, the patient should meet with a specialist in rehabilitation before surgery to understand what will be involved.

Amputation: Amputation is surgery to remove part or all of a limb (an arm or leg). When used to treat cancer, amputation removes the limb part with the tumor, some healthy tissue above it, and everything below it. In the past, amputation was the main way to treat bone cancers found in the arms or legs. Now, this operation is only chosen if there is a reason not to do limb-salvage surgery. For example, an amputation may be needed if removing all of the cancer requires removing essential nerves, arteries, or muscles that would leave the limb without good function.

MRI scans and examination of the tissue by the pathologist at the time of surgery can help the surgeon decide how much of the arm or leg needs to be removed. Surgery is planned so that muscles and the skin will form a cuff around the amputated bone. This cuff fits into the end of an artificial limb (or prosthesis). After surgery, the patient must learn how to use the prosthesis in rehabilitation. With proper physical therapy the patient is often walking again 3 to 6 months after leg amputation.

Limb-salvage surgery: The goal of limb-salvage surgery is to remove all of the cancer and still leave the patient with a working leg (or arm). Over 90% of patients with bone cancer in a limb are able to have their limb spared. This type of surgery is very complex and requires

surgeons with special skills and experience. The challenge for the surgeon is to remove the entire tumor while still saving the nearby tendons, nerves, and vessels. This is not always possible. If a cancer has grown into these structures, they will need to be removed along with the tumor. This can sometimes result in a limb that is painful or can't be used. In that case, amputation may be the best option.

In this type of surgery, a wide-excision is done to remove the tumor. A bone graft or an *endoprosthesis* (meaning internal prosthesis) is used to replace the bone that is lost. Endoprostheses are made of metal and other materials and can be very sophisticated. Because they may be used in growing children, some can be made longer without any extra surgery as the child grows.

Further surgery could be needed if the bone graft or endoprosthesis becomes infected, loose, or broken. Limb-salvage surgery patients may need more surgery during the following 5 years, and some may eventually need an amputation.

Rehab is much more intense after limb-salvage surgery than it is after amputation. It takes an average time of a year for patients to learn to walk again after limb-salvage of a leg. If the patient does not take part in the rehabilitation program, the salvaged arm or leg may become useless.

Reconstructive surgery: If the leg must be amputated mid-thigh, the lower leg and foot can be rotated and attached to the thigh bone. The old ankle joint becomes the new knee joint. This surgery is called *rotationplasty*. A prosthesis is used to make the new leg the same length as the other (healthy) leg.

If the bone tumor is located in the upper arm, the tumor may be removed and then the lower arm attached again. This leaves the patient with an arm that works but is much shorter.

Tumors in other areas

Bone cancer in the pelvis is treated with a wide-excision when possible. If needed, bone grafts can be used to rebuild the pelvic bones.

For a tumor in the lower jaw bone, the entire lower half of the jaw may be removed and later replaced with bones from other parts of the body.

For tumors in areas like the spine or the skull, it may not be possible to safely do a wide-excision. Cancers in these bones may require a combination of treatments such as curettage, cryosurgery, and radiation.

Curettage: This procedure involves scooping out the tumor from the bone without removing a section of the bone. This leaves a hole in the bone. In some cases, after most of the tumor has been removed, the surgeon will treat the nearby bone tissue to kill any remaining tumor cells. This can be done with cryosurgery or by using bone cement.

Cryosurgery: For this treatment, liquid nitrogen is poured into the hole that is left in the bone after the tumor was removed. This extremely cold material kills tumor cells by freezing them. This treatment is also called *cryotherapy*. After cryosurgery, the hole in the bone can be filled by bone grafts or by bone cement.

Bone cement: The bone cement polymethylmethyacrylate (PMMA) starts out as a liquid and hardens over time. It can be put into a hole in the bone in liquid form. As PMMA hardens, it gives off a lot of heat. The heat helps kill any remaining tumor cells. This allows PMMA to be used without cryosurgery for some types of bone tumors.

Surgical treatment of metastasis

To be able to cure a bone cancer, it and any existing metastases must be removed completely with surgery. The lungs are the most common site of distant spread for bone cancer. Surgery to remove bone cancer metastases to the lungs must be planned very carefully. Before the operation, the surgeon will consider the number of tumors, their location (one lung or both lungs), their size, and the patient's general condition.

The chest CT scan may not show all the tumors that truly exist. The surgeon will have a treatment plan in case more tumors are found during the operation than can be seen in the chest CT scan.

Removing all the lung metastases likely gives the patient the only chance for cure. However, not all lung metastases can be removed. Some tumors are too big or are too close to important structures in the chest (such as large blood vessels) to be removed safely. Patients whose general condition is not good (due to poor nutritional status or problems with the heart, liver, or kidneys) may not be able to withstand the stress of anesthesia and surgery to remove metastases.

Radiation therapy for bone cancer

Radiation therapy uses high-energy rays or particles to kill cancer cells. *External beam radiation therapy* uses radiation delivered from outside the body that is focused on the cancer. This is the type of radiation therapy that has been tried as a treatment for bone cancer.

Most bone cancers are not easily killed by radiation, and high doses are needed. These high doses can damage healthy structures like nerves in the nearby area. This is why radiation therapy does not play a major role in treating most types of bone tumors, (except for Ewing tumors, and these are discussed in a separate document). Often radiation is used to treat bone cancers that are *unresectable* (they cannot be completely removed by surgery). Radiation may also be used after surgery if cancer cells were present in the edges of the removed tissue. Another term for this is *positive margins* (this was discussed in the section about surgery). In this case, radiation may be given to kill any cancer that may have been left behind. If the cancer comes back after treatment, radiation can help control symptoms like pain and swelling.

Intensity-modulated radiation therapy

Intensity-modulated radiation therapy (IMRT) is an advanced form of external beam radiation therapy. With this technique, a computer matches the radiation beams to the shape of the tumor and can adjust the intensity (strength) of the beams. The radiation is delivered to the tumor from several directions to reduce the amount of radiation that goes through any one area of normal tissue. Altogether, this makes it possible to reduce radiation damage to normal tissues while increasing the radiation dose to the cancer.

Proton-beam radiation

Proton-beam radiation is a special form of radiation that uses protons instead of regular x-rays to kill cancer cells. Protons are positively charged particles that are found inside all atoms. They cause little damage to the tissues they pass through but are very good at killing cells at the end of their path. This allows a high dose of radiation to be given to the tumor without hurting the normal tissue around it. Proton-beam radiation therapy requires highly specialized equipment and is not available in all medical centers. This form of radiation has been found to be very helpful in treating skull base chondrosarcomas and chordomas.

Chemotherapy for bone cancer

Chemotherapy (chemo) is the use of drugs to treat cancer. Chemo is *systemic* therapy. This means that the drug enters the bloodstream and circulates to reach and destroy the cancer cells throughout the body.

Chemo is often a part of treatment for Ewing sarcoma and osteosarcoma, but it isn't often used for other bone cancers, like chordomas and chondrosarcomas, because they aren't very sensitive to chemo and so it often doesn't work well. It can be useful for some special types of chondrosarcoma, like *dedifferentiated* and *mesenchymal*.

Chemo is sometimes used for bone cancer that has spread through the bloodstream to the lungs and/or other organs. The drugs mainly used to treat bone cancer include:

- Doxorubicin (Adriamycin®)
- Cisplatin
- Carboplatin
- Etoposide (VP-16)
- Ifosfamide (Ifex®)
- Cyclophosphamide (Cytoxan®)
- Methotrexate

- Vincristine (Oncovin®)

Usually, several drugs (2 or 3) are given together. For example, a very common combination is cisplatin and doxorubicin. Other combinations are ifosfamide and etoposide or ifosfamide and doxorubicin

Side effects of chemotherapy

Chemo kills cancer cells, but it will also damage some normal cells. Careful attention is given to avoid or minimize side effects. The side effects of chemo depend on the type of drugs, the amount taken, and the length of time they are taken.

Some common temporary side effects can include:

- Nausea and vomiting
- Loss of appetite
- Hair loss
- Mouth sores

It is important to tell your cancer care team about any side effects you have so that they can be prevented or controlled.

Chemotherapy can damage the blood-producing cells of the bone marrow and lymph nodes, so patients may have low blood cell counts. Low blood cell counts can result in:

- Increased chance of infection (too few white blood cells)
- Bleeding or bruising after minor cuts or injuries (too few platelets)
- Fatigue or shortness of breath (too few red blood cells)

Some side effects are specific to particular drugs. It's important to note that many of the serious side effects are rare, but possible. Discuss these with your cancer care team if you have concerns before treatment.

Ifosfamide and cyclophosphamide can cause bloody urine by damaging the lining of the bladder. This is called *hemorrhagic cystitis.* This problem can be prevented by giving a drug called *mesna* along with the chemo.

Cisplatin may cause nerve damage (called *neuropathy*) leading to problems with numbness, tingling, and even pain in the hands and feet. Kidney damage (called *nephropathy*) can also occur after treatment with cisplatin. Giving lots of fluid before and after the drug is infused can help prevent this side effect. Cisplatin can sometimes cause problems with hearing (known as *ototoxicity*). Most often patients with this problem notice problems hearing high-pitched sounds.

Over time, doxorubicin can damage the heart. The risk of this goes up as the total amount of the drug given goes up. Before giving doxorubicin, your doctor may order a test of your heart function to make sure that it is safe to give you this drug.

The doctors and nurses will watch closely for side effects. There are treatments for most side effects, but preventing significant side effects is more important. Most, if not all, of these side effects will eventually stop after the treatment is over. Do not hesitate to ask any questions about side effects with the cancer care team.

While you are being treated, your doctor will order lab tests to be sure your liver, kidney, and bone marrow (which produces the cells in the blood) are functioning well.

- The **complete blood count (CBC)** includes levels of white blood cells, red blood cells, and platelets.
- **Chemistry panels** measure certain blood chemicals that tell doctors how well the liver and the kidneys are working. Some drugs used in chemotherapy can damage the kidneys and liver.

If one of the drugs that is being given can damage hearing, the doctor may order a hearing test (called an *audiogram*) before starting chemotherapy.

Targeted therapy for bone cancer

As researchers have learned more about the molecular and genetic changes in cells that cause cancer, they have been able to develop newer drugs that specifically target some of these changes. These drugs, often called *targeted therapy drugs*, work differently from standard chemotherapy (chemo) drugs and have different side effects. Targeted drugs are proving to be especially important in diseases such as chordomas and other bone cancers, where chemo has not been very useful.

Some chordomas have gene defects (mutations) that make proteins that signal the cells to grow. These genes are called *c-kit*, *PDGFRA*, and *PDGFRB*. The drug imatinib (Gleevec®) is a targeted therapy drug that can block the signals from these genes. This can cause some tumors to stop growing or even shrink a little. Imatinib is used to treat chordomas that have spread or have come back after treatment. Imatinib has been used to treat chordoma for several years, but it isn't approved by the Food and Drug Administration to treat this type of cancer. It is approved to treat more common cancers.

Clinical trials for bone cancer

You may have had to make a lot of decisions since you've been told you have cancer. One of the most important decisions you will make is choosing which treatment is best for you. You may have heard about clinical trials being done for your type of cancer. Or maybe someone on your health care team has mentioned a clinical trial to you.

Clinical trials are carefully controlled research studies that are done with patients who volunteer for them. They are done to get a closer look at promising new treatments or procedures.

If you would like to take part in a clinical trial, you should start by asking your doctor if your clinic or hospital conducts clinical trials. You can also call our clinical trials matching service for a list of clinical trials that meet your medical needs. You can reach this service at 1-800-303-5691 or on our Web site at www.cancer.org/clinicaltrials. You can also get a list of current clinical trials by calling the National Cancer Institute's Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237) or by visiting the NCI clinical trials Web site at www.cancer.gov/clinicaltrials.

There are requirements you must meet to take part in any clinical trial. If you do qualify for a clinical trial, it is up to you whether or not to enter (enroll in) it.

Clinical trials are one way to get state-of-the art cancer treatment. They are the only way for doctors to learn better methods to treat cancer. Still, they are not right for everyone.

You can get a lot more information on clinical trials in our document called *Clinical Trials: What You Need to Know*. You can read it on our Web site or call our toll-free number (1-800-227-2345) and have it sent to you.

Complementary and alternative therapies for bone cancer

When you have cancer you are likely to hear about ways to treat your cancer or relieve symptoms that your doctor hasn't mentioned. Everyone from friends and family to Internet groups and Web sites offer ideas for what might help you. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

What exactly are complementary and alternative therapies?

Not everyone uses these terms the same way, and they are used to refer to many different methods, so it can be confusing. We use *complementary* to refer to treatments that are used *along with* your regular medical care. *Alternative* treatments are used *instead of* a doctor's medical treatment.

Complementary methods: Most complementary treatment methods are not offered as cures for cancer. Mainly, they are used to help you feel better. Some methods that are used along with regular treatment are meditation to reduce stress, acupuncture to help relieve pain, or peppermint tea to relieve nausea. Some complementary methods are known to help, while others have not been tested. Some have been proven not be helpful, and a few have even been found harmful.

Alternative treatments: Alternative treatments may be offered as cancer cures. These treatments have not been proven safe and effective in clinical trials. Some of these methods

may pose danger, or have life-threatening side effects. But the biggest danger in most cases is that you may lose the chance to be helped by standard medical treatment. Delays or interruptions in your medical treatments may give the cancer more time to grow and make it less likely that treatment will help.

Finding out more

It is easy to see why people with cancer think about alternative methods. You want to do all you can to fight the cancer, and the idea of a treatment with no side effects sounds great. Sometimes medical treatments like chemotherapy can be hard to take, or they may no longer be working. But the truth is that most of these alternative methods have not been tested and proven to work in treating cancer.

As you consider your options, here are 3 important steps you can take:

- Look for "red flags" that suggest fraud. Does the method promise to cure all or most cancers? Are you told not to have regular medical treatments? Is the treatment a "secret" that requires you to visit certain providers or travel to another country?
- Talk to your doctor or nurse about any method you are thinking about using.
- Contact us at 1-800-227-2345 to learn more about complementary and alternative methods in general and to find out about the specific methods you are looking at.

The choice is yours

Decisions about how to treat or manage your cancer are always yours to make. If you want to use a non-standard treatment, learn all you can about the method and talk to your doctor about it. With good information and the support of your health care team, you may be able to safely use the methods that can help you while avoiding those that could be harmful.

Treating specific bone cancers

For specific information on treating Ewing sarcoma and osteosarcoma, please see the American Cancer Society documents on those cancers.

Chondrosarcomas

After a biopsy confirms the diagnosis, surgery is done to remove the tumor. Again, it is important that the biopsy be done by the same surgeon who will remove the tumor. For a low-grade chondrosarcoma in an arm or leg, curettage with cryotherapy is an option. If the tumor is high-grade, limb-sparing surgery will be done if possible. Sometimes amputation is needed to completely remove the cancer. If the chondrosarcoma has spread to the lung and there are only a few metastases, they may be removed surgically.

Chondrosarcomas in the skull are hard to treat. Complete surgical removal is difficult, and may cause serious side effects. Some low-grade tumors are treated with curettage and cryosurgery. Sometimes the patient is treated with radiation therapy. Since chondrosarcomas are resistant to radiation, high doses are required. Proton-beam radiation works well for these tumors.

Chemotherapy (chemo) is not often used to treat chondrosarcoma, but it is used to treat some variants of this cancer. For example, dedifferentiated chondrosarcoma may be treated like osteosarcoma, with chemo followed by surgery and then more chemo. Patients with mesenchymal chondrosarcomas also receive chemo before surgery. These tumors are treated the same as Ewing tumors or soft tissue sarcomas.

Malignant fibrous histiocytomas (MFH)

MFH is treated the same way osteosarcoma is treated. (Please see our document, *Osteosarcoma* for more detailed information.) Often the patient is first treated with chemotherapy to shrink the tumor. Then a wide-resection is done to remove the tumor (see the previous section about treatment for more details). After resection, the bone may be reconstructed with a bone graft or a prosthesis (metallic rod). Amputation is rarely needed. In some cases, chemotherapy is also given after surgery.

Fibrosarcomas

Surgery is the main treatment for this kind of cancer, with the goal of removing the tumor and a margin of surrounding normal bone. Radiation is sometimes given after surgery if the doctor suspects that some cancer has been left behind. Radiation therapy is sometimes used instead of surgery if the tumor cannot be removed completely. Radiation is also used if a fibrosarcoma returns after surgery.

Giant cell tumors of bone

These are treated mainly with surgery. Different surgeries are used, depending on the size and location of the tumor. One option is to remove the part of the bone affected by the tumor, replacing it with a bone graft or prosthesis (such as a metal rod). If this operation can be done without seriously affecting the movement of the limb or without causing serious damage to nearby tissues, this approach provides a good likelihood of success.

One option to wide-excision is curettage followed by cryosurgery. The defect (hole) in the bone can then be filled in with bone cement or a bone graft. Radiation therapy may sometimes be used for giant cell tumors in bones where surgery may be difficult to perform without damaging nearby sensitive tissues − such as the skull and the spine. Radiation is not often used to treat giant cell tumors because if the tumor is not killed completely it may increase the chance that it comes back in the malignant form.

Amputation is rarely needed to treat a giant cell tumor.

If a giant cell bone tumor spreads to other organs, the lungs are most commonly affected. If there are only a few metastatic tumors in the lungs, it may be possible to remove them surgically. Metastases can also be treated by with radiation.

Chordomas

This primary tumor of bone most often occurs in the base of the skull or the bones of the spine. Removing the tumor completely with surgery is not always possible because the spinal cord and nerves nearby may be involved. Radiation is often given after surgery to lower the chance that the tumor will grow back. Proton-beam radiation, either alone or with intensity-modulated radiation therapy, is often used. Imatinib (Gleevec) is often used for a chordoma that has spread widely. It may rarely shrink the tumors, but often can stop them from growing for a while. Chemo may be tried as well, but so far it hasn't worked well. Chordomas can come back, even 10 or more years after treatment, so long-term follow-up is important.

More treatment information about bone cancer

For more details on treatment options − including some that may not be addressed in this document − the National Comprehensive Cancer Network (NCCN) and the National Cancer Institute (NCI) are good sources of information.

The NCCN, made up of experts from many of the nation's leading cancer centers, develops cancer treatment guidelines for doctors to use when treating patients. Those are available on the NCCN Web site (www.nccn.org).

The NCI provides treatment guidelines by telephone (1-800-4-CANCER) and on its Web site (www.cancer.gov). Detailed guidelines intended for use by cancer care professionals are also available on www.cancer.gov.

What should you ask your doctor about bone cancer?

As you cope with cancer and cancer treatment, you need to have honest, open discussions with your doctor. You should feel free to ask any question that's on your mind no matter how small it might seem. Nurses, social workers, and other members of the treatment team may also be able to answer many of your questions.

- What kind of bone cancer do I have?
- Has my cancer spread beyond the primary site?
- What is the stage of my cancer and what does that mean in my case?
- What treatment choices do I have?

- What do you recommend and why?
- What risks or side effects are there to the treatments you suggest?
- What are the chances of my cancer coming back with these treatment plans?
- What should I do to be ready for treatment?
- Based on what you've learned about my cancer, how long do you think I'll survive?

In addition to these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery times so that you can plan your work schedule. Or you may want to ask about second opinions or about clinical trials for which you may qualify.

What happens after treatment for bone cancer?

For some people with bone cancer, treatment may remove or destroy the cancer. Completing treatment can be both stressful and exciting. You may be relieved to finish treatment, but find it hard not to worry about cancer coming back. (When cancer comes back after treatment, it is called *recurrence*.) This is a very common concern in people who have had cancer.

It may take a while before your fears lessen. But it may help to know that many cancer survivors have learned to live with this uncertainty and are leading full lives. Our document, *Living with Uncertainty: The Fear of Cancer Recurrence*, gives more detailed information on this.

For other people, the cancer may never go away completely. These people may get regular treatments with chemotherapy, radiation therapy, or other therapies to try to help keep the cancer in check. Learning to live with cancer that does not go away can be difficult and very stressful. It has its own type of uncertainty. Our document, *When Cancer Doesn't Go Away*, talks more about this.

Follow-up care

When treatment ends, your doctors will still want to watch you closely. It is very important to go to all of your follow-up appointments. During these visits, your doctors will ask questions about any problems you might be having and could use exams and lab tests or x-rays and scans to look for signs of cancer or treatment side effects. Almost any cancer treatment can have side effects. Some may last for a few weeks to months, but others can last the rest of your life. Now is the time for you to talk to your cancer care team about any changes or problems you notice and any questions or concerns you have.

Following extensive bone surgery, a program of rehabilitation and physical therapy will be an important part of helping you regain as much of your mobility and independence as possible.

It is important to keep health insurance. Tests and doctor visits cost a lot, and even though no one wants to think of their cancer coming back, this could happen.

Should your cancer come back, our document, *When Your Cancer Comes Back: Cancer Recurrence* can give you information on how to manage and cope with this phase of your treatment.

Seeing a new doctor

At some point after your cancer diagnosis and treatment, you may find yourself seeing a new doctor who does not know anything about your medical history. It is important for you to be able to give your new doctor the details of your diagnosis and treatment. Gathering these details soon after treatment may be easier than trying to get them at some point in the future. Make sure you have the following information handy:

- A copy of your pathology report(s) from any biopsies or surgeries
- If you had surgery, a copy of your operative report(s)
- If you had radiation, a copy of the treatment summary
- If you were in the hospital, a copy of the discharge summary that doctors prepare when patients are sent home
- If you had chemotherapy (or targeted therapy), a list of your drugs, drug doses, and when you took them
- A copy of your x-rays and other imaging studies (these can be put on a CD or DVD)

The doctor may want copies of this information for his records, but always keep copies for yourself.

Lifestyle changes after treatment for bone cancer

You can't change the fact that you have had cancer. What you can change is how you live the rest of your life – making choices to help you stay healthy and feel as well as you can. This can be a time to look at your life in new ways. Maybe you are thinking about how to improve your health over the long term. Some people even start during cancer treatment.

Making healthier choices

For many people, a diagnosis of cancer helps them focus on their health in ways they may not have thought much about in the past. Are there things you could do that might make you healthier? Maybe you could try to eat better or get more exercise. Maybe you could cut down on the alcohol, or give up tobacco. Even things like keeping your stress level under control may help. Now is a good time to think about making changes that can have positive effects for the rest of your life. You will feel better and you will also be healthier.

You can start by working on those things that worry you most. Get help with those that are harder for you. For instance, if you are thinking about quitting smoking and need help, call the American Cancer Society for information and support. This tobacco cessation and coaching service can help increase your chances of quitting for good.

Eating better

Eating right can be hard for anyone, but it can get even tougher during and after cancer treatment. Treatment may change your sense of taste. Nausea can be a problem. You may not feel like eating and lose weight when you don't want to. Or you may have gained weight that you can't seem to lose. All of these things can be very frustrating.

If treatment caused weight changes or eating or taste problems, do the best you can and keep in mind that these problems usually get better over time. You may find it helps to eat small portions every 2 to 3 hours until you feel better. You may also want to ask your cancer team about seeing a dietitian, an expert in nutrition who can give you ideas on how to deal with these treatment side effects.

One of the best things you can do after cancer treatment is put healthy eating habits into place. You may be surprised at the long-term benefits of some simple changes, like increasing the variety of healthy foods you eat. Getting to and staying at a healthy weight, eating a healthy diet, and limiting your alcohol intake may lower your risk for a number of types of cancer, as well as having many other health benefits.

Rest, fatigue, and exercise

Extreme tiredness, called *fatigue*, is very common in people treated for cancer. This is not a normal tiredness, but a "bone-weary" exhaustion that doesn't get better with rest. For some people, fatigue lasts a long time after treatment, and can make it hard for them to exercise and do other things they want to do. But exercise can help reduce fatigue. Studies have shown that patients who follow an exercise program tailored to their personal needs feel better physically and emotionally and can cope better, too.

If you were sick and not very active during treatment, it is normal for your fitness, endurance, and muscle strength to decline. Any plan for physical activity should fit your own situation. An older person who has never exercised will not be able to take on the same

amount of exercise as a 20-year-old who plays tennis twice a week. If you haven't exercised in a few years, you will have to start slowly – maybe just by taking short walks.

Talk with your health care team before starting anything. Get their opinion about your exercise plans. Then, try to find an exercise buddy so you're not doing it alone. Having family or friends involved when starting a new exercise program can give you that extra boost of support to keep you going when the push just isn't there.

If you are very tired, you will need to balance activity with rest. It is OK to rest when you need to. Sometimes it's really hard for people to allow themselves to rest when they are used to working all day or taking care of a household, but this is not the time to push yourself too hard. Listen to your body and rest when you need to. (For more information on dealing with fatigue, please see *Fatigue in People With Cancer* and *Anemia in People With Cancer*.)

Keep in mind exercise can improve your physical and emotional health.

- It improves your cardiovascular (heart and circulation) fitness.
- Along with a good diet, it will help you get to and stay at a healthy weight.
- It makes your muscles stronger.
- It reduces fatigue and helps you have more energy.
- It can help lower anxiety and depression.
- It can make you feel happier.
- It helps you feel better about yourself.

And long term, we know that getting regular physical activity plays a role in helping to lower the risk of some cancers, as well as having other health benefits.

How does having bone cancer affect your emotional health?

When treatment ends, you may find yourself overcome with many different emotions. This happens to a lot of people. You may have been going through so much during treatment that you could only focus on getting through each day. Now it may feel like a lot of other issues are catching up with you.

You may find yourself thinking about death and dying. Or maybe you're more aware of the effect the cancer has on your family, friends, and career. You may take a new look at your relationship with those around you. Unexpected issues may also cause concern. For instance, as you feel better and have fewer doctor visits, you will see your health care team less often and have more time on your hands. These changes can make some people anxious.

Almost everyone who has been through cancer can benefit from getting some type of support. You need people you can turn to for strength and comfort. Support can come in

many forms: family, friends, cancer support groups, church or spiritual groups, online support communities, or one-on-one counselors. What's best for you depends on your situation and personality. Some people feel safe in peer-support groups or education groups. Others would rather talk in an informal setting, such as church. Others may feel more at ease talking one-on-one with a trusted friend or counselor. Whatever your source of strength or comfort, make sure you have a place to go with your concerns.

The cancer journey can feel very lonely. It is not necessary or good for you to try to deal with everything on your own. And your friends and family may feel shut out if you do not include them. Let them in, and let in anyone else who you feel may help. If you aren’t sure who can help, call your American Cancer Society at 1-800-227-2345 and we can put you in touch with a group or resource that may work for you.

If treatment for bone cancer stops working

If cancer keeps growing or comes back after one kind of treatment, it is possible that another treatment plan might still cure the cancer, or at least shrink it enough to help you live longer and feel better. But when a person has tried many different treatments and the cancer has not gotten any better, the cancer tends to become resistant to all treatment. If this happens, it's important to weigh the possible limited benefits of a new treatment against the possible downsides. Everyone has their own way of looking at this.

This is likely to be the hardest part of your battle with cancer − when you have been through many medical treatments and nothing's working anymore. Your doctor may offer you new options, but at some point you may need to consider that treatment is not likely to improve your health or change your outcome or survival.

If you want to continue to get treatment for as long as you can, you need to think about the odds of treatment having any benefit and how this compares to the possible risks and side effects. In many cases, your doctor can estimate how likely it is the cancer will respond to treatment you are considering. For instance, the doctor may say that more chemo or radiation might have about a 1% chance of working. Some people are still tempted to try this. But it is important to think about and understand your reasons for choosing this plan.

No matter what you decide to do, you need to feel as good as you can. Make sure you are asking for and getting treatment for any symptoms you might have, such as nausea or pain. This type of treatment is called *palliative care*.

Palliative care helps relieve symptoms, but is not expected to cure the disease. It can be given along with cancer treatment, or can even be cancer treatment. The difference is its purpose - the main purpose of palliative care is to improve the quality of your life, or help you feel as good as you can for as long as you can. Sometimes this means using drugs to help with symptoms like pain or nausea. Sometimes, though, the treatments used to control your symptoms are the same as those used to treat cancer. For instance, radiation might be used to help relieve bone pain caused by cancer that has spread to the bones. Or chemo might be

used to help shrink a tumor and keep it from blocking the bowels. But this is not the same as treatment to try to cure the cancer.

At some point, you may benefit from hospice care. This is special care that treats the person rather than the disease; it focuses on quality rather than length of life. Most of the time, it is given at home. Your cancer may be causing problems that need to be managed, and hospice focuses on your comfort. You should know that while getting hospice care often means the end of treatments such as chemo and radiation, it doesn't mean you can't have treatment for the problems caused by your cancer or other health conditions. In hospice the focus of your care is on living life as fully as possible and feeling as well as you can at this difficult time. You can learn more about hospice in our document called *Hospice Care*.

Staying hopeful is important, too. Your hope for a cure may not be as bright, but there is still hope for good times with family and friends − times that are filled with happiness and meaning. Pausing at this time in your cancer treatment gives you a chance to refocus on the most important things in your life. Now is the time to do some things you've always wanted to do and to stop doing the things you no longer want to do. Though the cancer may be beyond your control, there are still choices you can make.

What’s new in bone cancer research and treatment?

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

Chemotherapy

Some clinical trials are looking into ways to combine surgery, radiation therapy, and chemotherapy (chemo). Some are testing new chemo drugs.

Targeted therapy

Targeted therapy drugs work differently from standard chemo. These drugs target certain genes and proteins in cancer cells.

One example of targeted therapy is the drug imatinib (Gleevec), which targets certain proteins made by the cancer cells in chordomas. Adding another drug to imatinib, such as the targeted therapy drug sirolimus (Rapamune®) or the chemo drug cisplatin helps stop the growth of chordomas when imatinib stops working. Another drug, LBH589, is being studied in combination with imatanib to treat chordoma.

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

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

Denosumab (Xgeva®, Prolia®) is a drug that was originally approved to treat osteoporosis and cancer that has spread to bone. In studies of patients with giant cell tumors of bone that had either come back after surgery or could not be removed with surgery, denosumab helped shrink tumors in some patients. On the basis of these studies, it was recently approved for this use by the US Food and Drug Administration.

Radiation

Some studies are looking at the best ways to give radiation to treat bone cancers. For example, studies are comparing proton beam radiation with carbon ion radiation in treating chordomas and chondrosarcomas of the skull base.

Genetics

In addition to clinical trials, researchers are making progress in learning about the causes of bone tumors. For example, changes to a certain part of chromosome 6 have been found in chordomas. Hopefully more information about the DNA changes that cause bone cancers will eventually lead to treatments aimed at these gene defects.

Additional resources for bone cancer

More information from your American Cancer Society

Here is more information you might find helpful. You also can order free copies of our documents from our toll-free number, 1-800-227-2345, or read them on our Web site, www.cancer.org.

After Diagnosis: A Guide for Patients and Families (also available in Spanish)

Caring for the Patient With Cancer at Home: A Guide for Patients and Families (also available in Spanish)

Ewing Family of Tumors

Osteosarcoma

Pain Control: A Guide for People With Cancer and Their Families (also available in Spanish)

Understanding Chemotherapy: A Guide for Patients and Families (also available in Spanish)

Understanding Radiation Therapy: A Guide for Patients and Families (also available in Spanish)

When Cancer Doesn't Go Away

When Your Cancer Comes Back: Cancer Recurrence

Your American Cancer Society also has books that you might find helpful. Call us at 1-800-227-2345 or visit our bookstore online at cancer.org/bookstore to find out about costs or to place an order.

National organizations and Web sites*

In addition to the American Cancer Society, other sources of patient information and support include:

National Cancer Institute
Toll-free number: 1-800-422-6237 (1-800-4-CANCER)
TTY: 1-800-332-8615
Web site: www.cancer.gov

**Inclusion on this list does not imply endorsement by the American Cancer Society.*

No matter who you are, we can help. Contact us anytime, day or night, for information and support. Call us at **1-800-227-2345** or visit www.cancer.org.

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