



Sarcoma: Adult Soft Tissue Cancer

What is cancer?

The body is made up of trillions of living cells. Normal body cells grow, divide to make new cells, and die in an orderly way. 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. In most cases the cancer cells form a tumor. Cancer cells can also invade (grow into) other tissues, something that normal cells can't 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 is 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 abnormal cell does.

People can inherit damaged DNA, but most often the 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.

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 named (and treated) based on where it started. For example, prostate cancer that has spread to the bones is still prostate cancer, not bone cancer.

Different types of cancer can behave very differently. They grow at different rates and respond to different treatments. This 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 can't 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 rarely life threatening.

What is a soft tissue sarcoma?

A sarcoma is a type of cancer that develops from certain tissues, like bone or muscle. Bone and soft tissue sarcomas are the main types of sarcoma. Soft tissue sarcomas can develop from soft tissues like fat, muscle, nerves, fibrous tissues, blood vessels, or deep skin tissues. They can be found in any part of the body. Most of them develop in the arms or legs. They can also be found in the trunk, head and neck area, internal organs, and the area in back of the abdominal cavity (known as the *retroperitoneum*). Sarcomas are not common tumors, and most cancers are the type of tumors called *carcinomas*.

Sarcomas that most often start in bones, such as osteosarcomas, and sarcomas that more often occur in children, such as the *Ewing Family of Tumors* and *Rhabdomyosarcoma*, are discussed separately.

There are many types of soft tissue tumors, and not all of them are cancerous. When the term sarcoma is part of the name of a disease, it means the tumor is malignant (cancer). Some soft tissue tumors behave in way in between a cancer and a non-cancer. These are called *intermediate*.

There are about 50 different types of soft tissue sarcomas (not all are listed here), such as:

- Adult fibrosarcoma
- Alveolar soft-part sarcoma
- Angiosarcoma (includes hemangiosarcoma and lymphangiosarcoma)
- Clear cell sarcoma
- Desmoplastic small round cell tumor
- Epithelioid sarcoma
- Fibromyxoid sarcoma, low-grade
- Gastrointestinal stromal tumor (GIST): this is a type of sarcoma that develops in the digestive tract. It is covered in a separate document, *Gastrointestinal Stromal Tumor (GIST)*.

- Kaposi sarcoma: this is a type of sarcoma that develops from the cells lining lymph or blood vessels. It is covered in a separate document, *Kaposi Sarcoma*
- Liposarcoma (includes dedifferentiated, myxoid, and pleomorphic liposarcomas)
- Leiomyosarcoma
- Malignant mesenchymoma
- Malignant peripheral nerve sheath tumors (includes neurofibrosarcomas, neurogenic sarcomas, and malignant schwannomas)
- Myxofibrosarcoma, low-grade
- Rhabdomyosarcoma: this is the most common type of soft tissue sarcoma seen in children and is discussed in the separate document *Rhabdomyosarcoma*.
- Synovial sarcoma
- Undifferentiated pleomorphic sarcoma (previously known as malignant fibrous histiocytoma or MFH)

Many of these types are discussed in more detail later in this section. There are many other types of tumors called soft tissue sarcomas, but these are all quite rare.

Intermediate soft-tissue tumors include:

- Dermatofibrosarcoma protuberans
- Fibromatosis (also known as desmoid tumor, musculoaponeurotic fibromatosis, and aggressive fibromatosis)
- Hemangioendothelioma
- Infantile fibrosarcoma
- Solitary fibrous tumor

Types of soft tissue sarcomas

- **Adult fibrosarcoma** usually affects fibrous tissue in the legs, arms, or trunk. It is most common in people between the ages of 20 and 60, but can occur in people of any age, even in infants.
- **Alveolar soft-part sarcoma** is a rare cancer that mostly affects young adults. These tumors most commonly occur in legs.

- **Angiosarcoma** can develop either from blood vessels (*hemangiosarcomas*) or from lymph vessels (*lymphangiosarcomas*). These tumors sometimes start in a part of the body that has been treated with radiation. Angiosarcomas are sometimes seen in the breast after radiation therapy and in limbs with lymphedema.
- **Clear cell sarcoma** is a rare cancer that often develops in tendons of the arms or legs. Under the microscope, it has some features of malignant melanoma, a type of cancer that develops from pigment-producing skin cells. How cancers with these features start in parts of the body other than the skin is not known.
- **Desmoplastic small round cell tumor** is a rare sarcoma of adolescents and young adults, found most often in the abdomen.
- **Epithelioid sarcoma** most often develops in tissues under the skin of the hands, forearms, feet, or lower legs. Adolescents and young adults are often affected.
- **Fibromyxoid sarcoma, low-grade** is a slow growing cancer that most often develops as a painless growth in the trunk or arms and legs (particularly the thigh). It is more common in young to middle aged adults. It is also sometimes called an *Evans' tumor*.
- **Liposarcomas** are malignant tumors of fat tissue. They can develop anywhere in the body, but they most often develop in the thigh, behind the knee, and inside the back of the abdomen. They occur mostly in adults between 50 and 65 years old..
- **Malignant mesenchymoma** is a rare type of sarcoma that shows features of fibrosarcoma and features of at least 2 other types of sarcoma.
- **Malignant peripheral nerve sheath tumors** include neurofibrosarcomas, malignant schwannomas, and neurogenic sarcomas. These are sarcomas that develop from the cells that surround a nerve.
- **Myxofibrosarcomas, low-grade** are most often found in the arms and legs of elderly patients. They are most common in or just under the skin and there might be more than one tumor nodule.
- **Synovial sarcoma** is a malignant tumor of the tissue around joints. The most common locations are the hip, knee, ankle, and shoulder. This tumor is more common in children and young adults, but it can occur in older people.
- **Undifferentiated pleomorphic sarcoma**, previously known as *malignant fibrous histiocytoma* (MFH), is most often found in the arms or legs. Less often, it can start inside at the back of the abdomen (the retroperitoneum). This sarcoma is most common in older adults. Although it mostly tends to grow locally, it can spread to distant sites.

Intermediate soft tissue tumors

- **Dermatofibrosarcoma protuberans** is a slow-growing cancer of the fibrous tissue beneath the skin, usually in the trunk or limbs. It grows into nearby tissues but rarely spreads to distant sites.
- **Fibromatosis** is the name given to fibrous tissue tumor with features in between fibrosarcoma and benign tumors such as fibromas and superficial fibromatosis. They tend to grow slowly but, often, steadily. They are also called *desmoid tumors*, as well as the more scientific name *musculoaponeurotic fibromatosis* or just *aggressive fibromatosis*. They rarely, if ever, spread to distant sites, but they do cause problems by growing into nearby tissues. They can sometimes be fatal. Some doctors consider them a type of low-grade fibrosarcoma; but others believe they are a unique type of fibrous tissue tumors. Certain hormones, particularly estrogen, make some desmoid tumors grow. Anti-estrogen drugs are sometimes useful in treating desmoids that cannot be completely removed by surgery.
- **Hemangioendothelioma** is a blood vessel tumor that is considered a low-grade cancer (meaning it grows slowly and is slow to spread). It does grow into nearby tissues and sometimes can spread to distant parts of the body (metastasize). It may start in soft tissues or in internal organs, such as the liver or lungs.
- **Infantile fibrosarcoma** is the most common soft tissue sarcoma in children under one year of age. It tends to be slow-growing and is less likely to spread to other organs than adult fibrosarcomas.
- **Solitary fibrous tumors** are most often not cancerous (benign) but can be malignant. Some start in the thigh, underarm, and pelvis. They can also start in the tissue surrounding the lung (called the pleura). Many tumors that were once called hemangiopericytomas are now considered solitary fibrous tumors.

Benign soft-tissue tumors

Many benign (non-cancerous) tumors can occur in soft-tissue as well. These include:

- Elastofibromas, benign tumor of fibrous tissue
- Fibromas, benign tumor of fibrous tissue
- Fibrous histiocyctomas, benign tumor of fibrous tissue
- Glomus tumors, benign tumors that occur near blood vessels
- Granular cell tumors: usually benign tumors in adults that occur often in the tongue but can be found almost anywhere in the body.

- Hemangiomas, benign tumors of blood vessels
- Hibernomas, benign tumors of fat tissue
- Lipomas, very common benign tumors of fat tissue
- Leiomyomas, benign tumors of smooth muscle that can be found anywhere in the body but are very common in the walls of the uterus where they are known as *fibroids*.
- Lipoblastomas, benign fat tissue tumors most often seen in children
- Lymphangiomas, benign tumors of lymph vessels
- Myxoma is a benign tumor that usually is located in muscles but does not start from muscle cells.
- Neurofibromas, tumors of nerve tissue that are usually benign. Neurofibromas of large nerves, such as those in the upper arms or neck can become cancerous. Neurofibromas are very common in people with an inherited condition called neurofibromatosis (also called von Recklinghausen disease), and are much less common in people without this condition.
- Neuromas, benign tumor of nerves that can be painful
- PEComa is a family of tumors made up of abnormal cells called *perivascular epithelial cells*. Although most of these tumors are benign, some rare PEComas are malignant (cancer). The most common of these tumors are angiomyolipoma and lymphangiomyoma. Angiomyolipoma is a benign tumor that most often affects the kidney. Lymphangiomyomatosis (LAM) is a rare disease of women in which the many lymphangiomyoma tumors grow into the lung tissue and interfere with lung function.
- Rhabdomyomas benign tumors of skeletal and heart muscle
- Schwannomas (neurilemmomas), benign tumors of the cells that coat nerves
- Tenosynovial giant cell tumor (also called nodular tenosynovitis), a benign tumor of joint tissue

Spindle cell tumors

Spindle cell tumor and spindle cell sarcoma are descriptive names based on the long, narrow appearance of the cells under the microscope. Spindle cell tumor is not a specific diagnosis or a specific type of cancer. The tumor may be a sarcoma, or it can be *sarcomatoid* — meaning another type of tumor (like a carcinoma) that looks like a sarcoma under the microscope.

Tumor-like conditions of soft tissue

Some conditions of soft tissues are caused by inflammation or injury and can form a mass that looks like a soft tissue tumor. Unlike a true tumor, they do not come from a single abnormal cell, they have limited capacity to grow or spread to nearby tissues, and never spread through the bloodstream or lymph system. *Nodular fasciitis* and *myositis ossificans* are 2 examples which affect tissues under the skin and muscle tissues, respectively.

What are the key statistics about soft tissue sarcomas?

The American Cancer Society's estimates for soft tissue sarcomas in the United States for 2015 are (these statistics include both adults and children):

- About 11,930 new soft tissue sarcomas will be diagnosed (6,610 cases in males and 5,320 cases in females).
- 4,870 Americans (2,600 males and 2,270 females) are expected to die of soft tissue sarcomas.

The most common types of sarcoma in adults are undifferentiated pleomorphic sarcoma (previously called malignant fibrous histiocytoma), liposarcoma, and leiomyosarcoma. Certain types occur more often in certain areas of the body than others. For example, leiomyosarcomas are the most common abdominal sarcoma, while liposarcomas and undifferentiated pleomorphic sarcoma are most common in legs. But pathologists (doctors who specialize in diagnosing cancers by how they look under the microscope), may not always agree on the exact type of sarcoma. Sarcomas of uncertain type are very common.

What are the risk factors for soft tissue sarcomas?

A *risk factor* is anything that changes your chance of getting a disease like cancer. Different cancers have different risk factors. For example, unprotected exposure to strong sunlight is a risk factor for skin cancer. Smoking is a risk factor for cancers of the lung, and many other cancers. But risk factors don't tell us everything. Having a risk factor, or even several, doesn't mean that you will get the cancer. Also, many people get cancer without having a risk factor.

Scientists have found a few risk factors that make a person more likely to develop soft tissue sarcomas. These include:

- Radiation

- Certain family cancer syndromes
- A damaged lymph system
- Exposure to certain chemicals

Injury and lifestyle factors such as smoking, diet, and exercise are NOT linked to the risk of soft tissue sarcoma.

Radiation given to treat other cancers

Patients might develop sarcomas from radiation given to treat other cancers, like breast cancer or lymphoma. The sarcoma often starts in the area of the body that had been treated with radiation. The average time between the radiation treatments and the diagnosis of a sarcoma is about 10 years. Radiation exposure accounts for less than 5% of sarcomas.

Radiation therapy techniques have improved steadily over several decades. Treatments now target cancers more precisely, and more is known about selecting radiation doses. These advances are expected to reduce the number of cancers caused by radiation therapy. But because these cancers take so long to develop, the results of these changes may not be seen for a long time. Still, radiation therapy is used only when its benefits (improved survival rate and relief of symptoms) outweigh the risk of cancer and other complications. For more information, see our document *Second Cancers in Adults*.

Family cancer syndromes

Family cancer syndromes are disorders caused by gene defects (mutations) that people are born with (often inherited from a parent) that are linked to a high risk of getting certain cancers. Some family cancer syndromes increase a person's risk of developing soft tissue sarcomas.

Neurofibromatosis

Neurofibromatosis usually runs in families and is characterized by many benign tumors that form in nerves under the skin and in other parts of the body (neurofibromas). It is also known as *von Recklinghausen disease*. It is caused by a defect (mutation) in a gene called *NF1*. About 5% of people with neurofibromatosis will develop a malignant peripheral nerve sheath tumor in a neurofibroma.

Gardner syndrome

Gardner syndrome is a disease caused by defects in the *APC* gene. This syndrome is a type of familial adenomatous polyposis, and people with it get many polyps in the colon (and intestines) and have a high risk of getting colon cancer. It also causes problems outside the

colon including desmoid tumors (these were discussed in the section “What is a soft tissue sarcoma?”).

Li-Fraumeni syndrome

Li-Fraumeni syndrome is caused by inherited defects in the *TP53* gene. People affected by this syndrome have a high risk of cancer, such as breast cancer, brain tumors, and sarcomas. People with this syndrome are sensitive to the cancer-causing effects of radiation. If their cancer is treated with radiation, they have a very high chance of developing a new cancer in the part of the body that received the radiation.

Retinoblastoma

Retinoblastoma is an eye cancer in children that can be caused by defects in the *RBI* gene. Children with one of these gene defects also have a higher risk of developing bone or soft tissue sarcomas, especially if treated for cancer with radiation.

Werner syndrome

Werner syndrome is caused by defects in the *RECQL2* gene. Children with this syndrome have problems like those seen in the elderly. These include cataracts, skin changes, and clogged heart arteries (arteriosclerosis) which can lead to heart attacks. They also have an increased risk of cancer, including soft tissue sarcomas.

Gorlin syndrome

Gorlin syndrome, also called *nevroid basal cell carcinoma syndrome*, is caused by defects in the *PTCH1* gene. People with this syndrome have a high risk of developing many basal cell skin cancers. They also have an increased risk of getting fibrosarcoma and rhabdomyosarcoma.

Tuberous sclerosis

Tuberous sclerosis can be caused by a defect in the *TSC1* gene. It can also be caused by a defect in another gene: *TSC2*. People with this syndrome often have seizures and learning problems. They get benign tumors in many different organs. They also get kidney problems, often along with a kidney tumor called *angiomyolipoma*. People with tuberous sclerosis have an increased risk of getting rhabdomyosarcoma.

Damaged lymph system

Lymph is a clear fluid containing immune system cells that is carried throughout the body by a series of lymph vessels. These vessels connect lymph nodes (small bean-shaped collections

of immune system cells). When lymph nodes have been removed by surgery or damaged by radiation therapy, lymph fluid can build up and cause swelling. This is called *lymphedema*.

Lymphangiosarcoma (a malignant tumor that develops in lymph vessels) is a very rare complication of chronic lymphedema.

Chemicals

Exposure to vinyl chloride (a chemical used in making plastics) is a risk factor for developing sarcomas of the liver, but it has not been proven to cause soft tissue sarcomas. Arsenic has also been linked to a type of liver sarcoma but not soft tissue sarcoma. Exposure to dioxin and to herbicides that contain phenoxyacetic acid at high doses (such as might occur in people who work on farms) may also be risk factors, but this is not known for certain. There is no evidence that herbicides (weed killers) or insecticides, at levels encountered by the general public, cause sarcomas.

Injury

An injury is **not** a risk factor for developing sarcomas. But this issue has caused some confusion in the past. One reason is that an injured area might swell. That swelling could resemble a tumor but is not a true tumor. Also, when you are injured, the pain may draw your attention to the injured area. The area may be examined closely, and x-rays or other imaging studies may be obtained. This can make it more likely that any sarcoma that is present will be discovered, even though it may have been present for some time.

Do we know what causes soft tissue sarcomas?

Scientists still don't know exactly what causes most cases of soft tissue sarcoma, but they have found several risk factors that can make a person more likely to develop these cancers. And research has shown that some of these risk factors affect the DNA of cells in the soft tissues.

Researchers have made great progress in understanding how certain changes in DNA can cause normal cells to become cancerous. Our 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 the way we look.

The 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 for proteins that control when our cells grow and divide.

Certain 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 turn on oncogenes or turn off tumor suppressor genes.

Several family cancer syndromes have been found in which inherited DNA mutations cause a very high risk of developing breast, colon, kidney, eye, or other cancers. In some of these, there is also an increased risk of developing soft tissue sarcomas (these were noted in “What are the risk factors for soft tissue sarcomas?”) They are caused by defects (mutations) in genes that can be inherited from a parent. These gene defects can be found through testing. For more on this topic, see *Family Cancer Syndromes* and *Genetic Testing: What You Need to Know*.

DNA mutations in soft tissue sarcoma are common. They are usually acquired during life rather than having been inherited before birth. Acquired mutations may result from exposure to radiation or cancer-causing chemicals. In most sarcomas, they occur for no apparent reason.

Researchers still do not know why most soft tissue sarcomas develop in people who have no apparent risk factors.

Can soft tissue sarcomas be prevented?

The only way to prevent some soft tissue sarcomas is to avoid exposure to risk factors whenever possible. Most sarcomas, however, develop in people with no known risk factors, so there is no way known at this time to prevent most cases. And for people receiving radiation therapy, there is usually little choice.

Can soft tissue sarcomas be found early?

People who have a strong family history of sarcomas or who have had other cancers when they were young, might wish to discuss the benefits and disadvantages of genetic testing with their doctor. The test results should always be explained by a genetic counselor or a specially trained doctor who can interpret the results and advise high-risk patients about the need for early cancer detection tests.

Families with a history of certain inherited conditions (see, “What are the risk factors for soft tissue sarcomas?”) caused by defects in certain genes have an increased risk of developing soft tissue sarcomas. The mutated genes can be detected by genetic testing, so family members should discuss this option with their doctors. They should let their doctor know about any lumps or growths right away.

No screening tests and exams are recommended for people who have no family history of sarcoma or other sarcoma risk factors. For these people, the best approach to early detection

is to tell their health care professional about any unexplained lumps or growths or other symptoms that may be caused by a soft tissue sarcoma.

Signs and symptoms of soft tissue sarcomas

More than half of sarcomas begin in an arm or leg. Most people simply notice a **lump** that has grown over a period of time (weeks to months). Although the lump is often not painful, in some cases it will hurt.

When sarcomas grow in the back of the abdomen (the retroperitoneum), the symptoms they cause more often come from other problems. Sometimes the tumors cause pain. They may also cause blockage or bleeding of the stomach or bowels. They can grow large enough for the tumor to be felt in the abdomen. About 20% of sarcomas begin in the abdomen (stomach) area.

Sarcomas can also begin on the outside of the chest or abdomen (about 10%) or in the head or neck area (around 10%).

If you have any of the following problems, see a doctor right away:

- **A new lump or a lump that is growing (anywhere on your body)**
- **Abdominal pain that is getting worse**
- **Blood in your stool or vomit**
- **Black, tarry stools** (when bleeding happens in the stomach or bowels, the blood can turn black as it is digested, and it may make the stool very black and sticky)

These symptoms are more often caused by things other than sarcoma, but they still need to be checked out by a doctor.

How are soft tissue sarcomas diagnosed?

If you have signs or symptoms or other reasons to suggest you might have a sarcoma, your doctor will talk with you, examine you, and probably need to order some tests to find out if you have cancer.

Imaging tests

Some tests, such as a computed tomography (CT) scan or a magnetic resonance imaging (MRI) scan, are often done to look for the cause of symptoms and to find a tumor (such as a sarcoma). Other tests may be done after a sarcoma is diagnosed to look for cancer spread.

Plain x-ray

A regular x-ray of the area with the lump may be the first test ordered. A chest x-ray may be done after you are diagnosed to see if the sarcoma has spread to the lungs.

Computed tomography 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 will create multiple images of the part of your body being studied. A CT scan is often done if the doctor suspects a soft tissue sarcoma in the chest, abdomen, or the retroperitoneum (the of the abdomen) . This test is also used to see if the sarcoma has spread into the lungs, liver or other organs.

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

Before any pictures are taken, you might be asked to drink 1 to 2 pints of a liquid called *oral contrast*. This helps outline the intestine more clearly. You may also receive an IV (intravenous) line through which a different kind of contrast dye (IV contrast) is injected. This helps better outline structures in your body.

The IV contrast dye can also cause some flushing (redness and warm feeling). Some people are allergic and get hives or, rarely, have more serious reactions like trouble breathing and low blood pressure. Be sure to tell the doctor if you have ever had a reaction to any contrast material used for x-rays.

CT scans might be done to precisely guide a biopsy needle into a tumor inside the body — the chest or abdomen, for example. 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 sure the needle is within the mass.

Magnetic resonance imaging scans

Magnetic resonance imaging (MRI) scans use radio waves and strong magnets instead of x-rays to take pictures of the body. 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. A contrast material might be injected, just as with CT scans, but is used less often.

MRI scans are often part of the work-up of any tumor that could be a sarcoma. They are often better than CT scans in evaluating sarcomas in the arms or legs. They provide a good picture of the extent of the tumor. They can show your health care team many things about the tumor, including location, size, and sometimes even the type of tissue it comes from (like fat or muscle). This makes MRI scans useful in planning a biopsy.

MRIs are also very helpful in examining 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 lie inside a long tube, which is confining and can be upsetting. Special "open" MRI machines sometimes are an option for people who have claustrophobia (fear of enclosed spaces), but the drawback is that the pictures are often not as clear. MRI machines also make a thumping noise that you may find disturbing. Some places will provide headphones with music to block this noise out.

Ultrasound

Ultrasound uses sound waves and their echoes to produce pictures of parts of the body. A small instrument called a *transducer* emits sound waves and picks up the echoes as they bounce off the organs. A computer converts the sound wave echoes into an image that is displayed on a computer screen.

This is a very easy procedure to have. It uses no radiation, which is why it is often used to look at developing fetuses. For most ultrasounds, you simply lie on a table while a technician moves the transducer over the part of your body being examined. Usually, the skin is first lubricated with gel. Ultrasound may be done before a biopsy to see if a lump is a cyst, meaning if it has fluid and is likely benign, or if it is solid and more likely a tumor. This test is often not needed if a CT or MRI was done.

Positron emission tomography scan

In this test, radioactive glucose (sugar) is injected into the patient's vein to look for cancer cells. Because cancers use glucose (sugar) at a higher rate than normal tissues, the radioactivity will tend to concentrate in the cancer. A scanner can spot the radioactive deposits. A positron emission tomography (PET) scan is useful when your doctor thinks the cancer has spread but doesn't know where. A PET scan can be used instead of several different x-rays because it scans your whole body. Often the PET scan is used with a CT scan. This helps decide if abnormalities seen on the CT scan are cancer or something else. PET is not often used for sarcoma, but it can be helpful in certain cases.

Biopsy

A biopsy is a procedure that removes a sample of tissue from a tumor to see if it is cancer. The piece of tissue is looked at under a microscope and, some other tests may be done on the sample as well. A physical exam or imaging test may suggest that a tumor is a sarcoma, but a

biopsy is the only way to be certain that it is a sarcoma and not another type of cancer or a benign disease.

Several types of biopsies are used to diagnose sarcomas. Doctors experienced with these tumors will choose one, based on the size and location of the tumor. Most prefer to use a fine needle aspiration or a core needle biopsy as the first step.

Fine needle aspiration (FNA) biopsy

In FNA, the doctor uses a very thin needle and a syringe to withdraw small pieces of tissue from the tumor mass. The doctor can often aim the needle while feeling the mass near the surface of the body. If the tumor is too deep to feel, the doctor can guide the needle while viewing it on a computed tomography (CT) scan or ultrasound. The main advantage of FNA is that it can be used to biopsy tumors deep in the body without surgery. The disadvantage is that the thin needle may not remove enough tissue to make a precise diagnosis.

FNA is often useful in showing that a mass first thought to be a sarcoma (found on physical exam or imaging tests) is really another type of cancer, a benign tumor, an infection, or some other disease. But if FNA results suggest a sarcoma, another type of biopsy will usually be done to remove enough tissue to confirm that diagnosis. After a sarcoma is diagnosed, FNA is most useful in determining whether additional tumors in other organs are metastases.

Core needle biopsy

Core needle biopsies use a needle that is larger than the FNA needle. Sometimes this needle is called a *Tru-Cut* needle. It removes a cylindrical piece of tissue about 1/16 inch across and 1/2 inch long. It usually removes enough tissue to see if a sarcoma is present. Like FNA, CT scan and ultrasound can be used to guide the needle into tumors of internal organs.

Surgical biopsy

In a surgical biopsy, the entire tumor or a piece of the tumor is removed during an operation. There are 2 types of surgical biopsies, excisional and incisional. In an excisional biopsy, the surgeon removes the entire tumor. In an incisional biopsy, only a piece of a large tumor is removed. An incisional biopsy almost always removes enough tissue to diagnose the exact type and grade of sarcoma. If the tumor is near the skin surface, this is a simple operation that can be done with local or regional anesthesia (numbing medication given near the mass or into a nerve). But if the tumor is deep inside the body, general anesthesia is used (the patient is asleep).

If a tumor is rather small, near the surface of the body, and not located near critical tissues (such as important nerves or large blood vessels), the doctor may choose to remove the entire mass and a margin of normal tissue in an excisional biopsy. This surgery combines the biopsy and the treatment into one operation, so it should only be done by a surgeon with experience in treating sarcomas.

If the tumor is large, then an incisional biopsy is needed. Only a surgeon experienced in sarcoma treatment should perform this procedure.

You might want to ask about your surgeon's experience with this procedure. Proper biopsy technique is a very important part of successfully treating soft tissue sarcomas. An improper biopsy can lead to tumor spread and problems removing the tumor later on. An incisional biopsy in the wrong place or an excision without wide enough margins can make it harder to completely remove a sarcoma later on. To prevent these problems, these 2 types of biopsies should only be done by a surgeon experienced in treating sarcomas. It is best that an incisional biopsy be done by the same surgeon who will later remove the entire tumor (if a sarcoma is found).

Testing biopsy samples

The tissue removed will be looked at under the microscope to see if cancer is present. If it is, the doctor will try to determine what kind it is (sarcoma or carcinoma).

Grading: If a sarcoma is present, the biopsy will be used to determine what type it is and its grade. The grade of a sarcoma is based on how the cancer cells look under the microscope. In grading a cancer, the pathologist (a doctor who specializes in diagnosing diseases by looking at the tissue under a microscope) considers how closely the tumor resembles normal tissue (*differentiation*), how many of the cells appear to be dividing, and how much of the tumor is made up of dying tissue.

Each factor is given a score, and the scores are added to determine the grade of the tumor. Sarcomas that have cells that look more normal and have fewer cells dividing are generally placed in a low-grade category. Low-grade tumors tend to be slow growing, slower to spread, and often have a better outlook (prognosis) than higher-grade tumors. Certain types of sarcoma are automatically given higher differentiation scores. This affects the overall score so much that they are never considered low grade. Examples of these include synovial sarcomas and embryonal sarcomas.

The grade is partly used to determine the stage of a sarcoma. The official staging system (see the section, "How are soft tissue sarcomas staged?") divides sarcomas into 3 grades (1 to 3). The grade of a sarcoma helps predict how rapidly it will grow and spread. It is useful in predicting a patient's outlook and helps determine treatment options.

Immunohistochemistry: Sometimes these special tests are needed to accurately determine whether a sarcoma is present and, if so, what type. Part of the biopsy sample is treated with special man-made antibodies that recognize cell proteins typical of certain kinds of sarcomas. The cells are treated with chemicals that make the cells containing these specific proteins change color. The color change is then seen under a microscope.

Cytogenetics: For this test, cells' chromosomes are examined with a microscope to look for changes. For example, in certain types of sarcomas part of one chromosome may be abnormally attached to part of a different chromosome (called a *translocation*). To see the

chromosomes clearly, the cancer cells must be grown in laboratory dishes until they start dividing. This can take a week or more.

Fluorescent in situ hybridization (FISH) can sometimes be used to detect translocations and other chromosome changes without first growing the cells in the lab. Tests of chromosome changes are not required to diagnose a sarcoma, but they are sometimes very useful in confirming that a certain type of sarcoma is present. And as new changes are discovered, these tests may become more important and more common.

Reverse transcription polymerase chain reaction (RT-PCR): This test is another way to find translocations in some sarcomas (such as the Ewing family of tumors, alveolar rhabdomyosarcoma, and synovial sarcoma) to confirm the type of tumor. Instead of using a microscope to look for the chromosome changes as in cytogenetic testing or FISH, RT-PCR uses chemical analysis of the RNA (a substance that is made from DNA) from genes affected by the translocation. RT-PCR testing is often able to find translocations that aren't detected by cytogenetics.

How are soft tissue sarcomas staged?

The process of finding out how far a cancer has spread is called *staging*. In sarcoma staging, doctors also evaluate the appearance of the tumor under the microscope and judge how fast the cancer seems to be growing. The stage of a sarcoma is the most significant factor in determining each patient's prognosis (the course of the disease and the chances of survival) and in selecting treatment options.

The information needed to stage sarcomas includes biopsies, and imaging tests of the main tumor (usually with CT or MRI scans) other parts of the body where the cancer may have spread.

When examining the biopsy sample, the pathologist (doctor who specializes in diagnosing diseases by looking at the tissue under a microscope) takes into account the number of cells that are actively dividing and how closely the cancer resembles normal tissue. He or she determines the cell type and grade and estimates how rapidly it will grow and spread.

A staging system is a standard way for the cancer care team to summarize the extent of a cancer's spread. The system often used to stage sarcomas is the TNM system of American Joint Committee on Cancer.

- **T** stands for the size of the **tumor**.
- **N** stands for spread to lymph **nodes** (small bean-shaped collections of immune system cells found throughout the body that help fight infections and cancers).
- **M** is for **metastasis** (spread to distant organs).

In soft tissue sarcomas, an additional factor, called grade (**G**), is also part of the tumor's stage. The grade is based on how the sarcoma cells look under the microscope.

Grade (G)

The grade is a sign of how likely it is the cancer will spread. Previously, the grade of a sarcoma was only based on how normal the cells looked under the microscope (called *differentiation*). This was not very helpful, and under a new system (known as the *French* or *FNCLCC system*), grade is based on 3 factors:

- **Differentiation:** Cancer cells are given a score of 1 to 3, with 1 being assigned when they look similar to normal cells and 3 being used when the cancer cells look very abnormal. Certain types of sarcoma are given a higher score automatically.
- **Mitotic count:** How many cancer cells are seen dividing under the microscope; given a score from 1 to 3 (a lower score means fewer cells were seen dividing)
- **Tumor necrosis:** How much of the tumor is made up of dying tissue; given a score from 0 to 2 (a lower score means there was less dying tissue present).

The scores for each factor are added to determine the grade for the cancer. Higher-grade cancers tend to grow and spread faster than lower-grade cancers.

GX: The grade cannot be assessed (because of incomplete information).

Grade 1 (G1): Total score of 2 or 3

Grade 2 (G2): Total score of 4 or 5

Grade 3 (G3): Total score of 6 or higher

Tumor (T)

T1: The sarcoma is 5 cm (2 inches) or less across

- **T1a:** The tumor is superficial (near the surface of the body).
- **T1b:** The tumor is deep in the limb or abdomen.

T2: The sarcoma is greater than 5 cm (2 inches) across.

- **T2a:** The tumor is superficial (near the surface of the body).
- **T2b:** The tumor is deep in the limb or abdomen.

Lymph nodes (N)

N0: The sarcoma has not spread to nearby lymph nodes.

N1: The sarcoma has spread to nearby lymph nodes.

Metastasis (M)

M0: No distant metastases (spread) of sarcoma are found.

M1: The sarcoma has spread to distant organs or tissues (such as the lungs).

Stage grouping for soft tissue sarcomas

To assign a stage, information about the tumor, its grade, lymph nodes, and metastasis is combined by a process called *stage grouping*. The stage is described by Roman numerals from I to IV and the letters A or B. The stage is useful in selecting treatment, but other factors, such as where the sarcoma is located, also influence treatment planning and outlook.

Stage IA

T1, N0, M0, G1 or GX: The tumor is not larger than 5 cm (2 inches) across (T1). It has not spread to lymph nodes (N0) or more distant sites (M0). The cancer is grade 1 (or the grade cannot be assessed).

Stage IB

T2, N0, M0, G1 or GX: The tumor is larger than 5 cm (2 inches) across (T2). It has not spread to lymph nodes (N0) or more distant sites (M0). The cancer is grade 1 (or the grade cannot be assessed).

Stage IIA

T1, N0, M0, G2 or G3: The tumor is not larger than 5 cm (2 inches) across (T1). It has not spread to lymph nodes (N0) or more distant sites (M0). The cancer is grade 2 or 3.

Stage IIB

T2, N0, M0, G2: The tumor is larger than 5 cm (2 inches) across (T2). It has not spread to lymph nodes (N0) or more distant sites (M0). The cancer is grade 2.

Stage III

Either

T2, N0, M0, G3: It is larger than 5 cm (2 inches) across (T2). It has not spread to lymph nodes (N0) or more distant sites (M0). The cancer is grade 3.

OR

Any T, N1, M0, any G: The cancer can be any size (any T) and any grade (any G). It has spread to nearby lymph nodes (N1). It has not spread to distant sites (M0).

Stage IV

Any T, Any N, M1, any G: The cancer can be any size (any T) and grade (any G). It has spread to lymph nodes near the tumor (N1) and/or to distant sites (M1).

Survival by stage of soft tissue sarcoma

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 don't want to know them, stop reading here and go to the next section.

The 5-year survival rate (or *observed* 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 effect of the cancer on survival.

To get 5-year survival rates, doctors have to look at people who were treated at least 5 years ago. If treatment has improved since then, people now being diagnosed with soft tissue sarcoma may have a more favorable outlook.

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 individual's case. Many other factors might affect a person's outlook, like the type of sarcoma, the location of the tumor, the treatment received, and the age of the patient. For example, sarcomas of the arms or legs have a better outcome than those found in other places. Also, older patients tend to have worse outcomes than younger people. Your doctor can tell you how the numbers below may apply to you, as he or she is familiar with your particular situation.

The rates below are based on the stage of the cancer *at the time of diagnosis*. When looking at survival rates, it's important to understand that the stage of a cancer does not change over time, even if the cancer progresses. A cancer that comes back or spreads is still referred to by the stage it was given when it was first found and diagnosed, but more information is added to explain the current extent of the cancer. (And the treatment plan is adjusted based on the change in cancer status.)

The overall relative 5-year survival rate of people with soft tissue sarcomas is around 50% according to statistics from the National Cancer Institute (NCI). These statistics include people with Kaposi sarcoma, which has a poorer outlook than many sarcomas. The NCI doesn't use the AJCC staging system. Instead, they group sarcomas only by whether they are still confined to the primary site (called *localized*) have spread to nearby lymph nodes or tissues (called *regional*); or have spread (metastasized) to sites away from the main tumor (called *distant*). The 5-year survival rates for soft tissue sarcomas have not changed much for many years. The corresponding 5-year relative survival rates were:

- 83% for localized sarcomas (56% of soft tissue sarcomas were localized when they were diagnosed)
- 54% for regional stage sarcomas; (19% were in this stage)
- 16% for sarcomas with distant spread (16% were in this stage)

The 10-year relative survival rate is only slightly worse for these stages, meaning that most people who survive 5 years are probably cured.

For sarcomas of the arms and legs, Memorial Sloan-Kettering Cancer Center has survival rates broken down by AJCC stage (these are for observed, not relative survival):

Stage	5-year observed survival rate
I	90%
II	81%
III	56%
IV	Not available

Survival is worse when the sarcoma has developed somewhere other than the arms or legs. For example, the 5-year survival for retroperitoneal sarcomas is around 40% to 60%.

How are soft tissue sarcomas 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

Experts recommend that patients with sarcoma have a health care team made up of doctors from different specialties, such as:

- An orthopedic surgeon: a surgeon who specializes in diseases of the bones, muscles, and joints (for sarcomas of the arms and legs)
- A surgical oncologist: a doctor who treats cancer with surgery (for sarcomas of the abdomen and retroperitoneum [the back of the abdomen])
- A thoracic surgeon: a doctor who treats diseases of the lungs and chest with surgery (for sarcomas in the chest)
- A medical oncologist: a doctor who treats cancer with medicines such as chemotherapy
- A radiation oncologist: a doctor who treats cancer with radiation therapy
- A physiatrist (or rehabilitation doctor): a doctor who treat injuries or illnesses that affect how you move

Many other specialists may be involved in your care as well, including physician assistants, nurse practitioners, nurses, respiratory therapists, social workers, physical therapists, and other health professionals.

After a sarcoma is found and staged, your team will recommend one or several treatment options. This decision is important, so take time and think about all of the choices. In choosing a treatment plan, factors to consider include the type, location, and stage of the cancer, as well as your overall physical health.

The main types of treatment for soft tissue sarcoma are:

- Surgery
- Radiation
- Chemotherapy
- Targeted therapy

It is important to discuss all of your treatment options, including their goals and possible side effects, with your doctors to help make the decision that best fits your needs. It's also very

important to ask questions if there is anything you're not sure about. You can find some good questions to ask in the section, "What should you ask your doctor about soft tissue sarcomas?" It also is often a good idea to seek a second opinion. A second opinion can provide more information and help you feel more confident about the chosen treatment plan. Some insurance companies require a second opinion before they will agree to pay for treatments.

Surgery for soft tissue sarcomas

Depending on the site and stage of a sarcoma, surgery might be able to remove the cancer and some of the nearby tissue. The goal of surgery is to remove the entire tumor along with at least 1 to 2 cm (less than an inch) of the normal tissue surrounding the tumor. This is to make sure that no cancer cells are left behind. When the removed tissue is looked at under a microscope, the doctor will check to see if cancer is growing in the edges (*margins*) of the specimen. If cancer cells are present at the edges, the tissue removed is said to have *positive margins*. This means that cancer cells may have been left behind.

When cancer cells are left after surgery, the patient may need more treatment – such as radiation or another surgery. If cancer isn't growing into the edges of the tissue removed, it is said to have *negative* or *clear margins*. The sarcoma has much less chance of coming back after surgery if it is removed with clear margins. When the tumor is in the abdomen, removing the tumor with enough normal tissue to get clear margins could be difficult because the tumor could be next to vital organs that can't be taken out.

In the past, many of the sarcomas in the arms and legs were treated by removing the limb (amputation). Now, this rarely is needed. Instead, most patients can be treated with surgery to remove the tumor without amputation (called *limb-sparing surgery*). This is usually followed by radiation therapy. These patients have the same survival rates as those who have amputations.

Sometimes, an amputation can't be avoided. It might be the only way to remove all of the cancer. Other times, critical nerves, muscles, bone, and blood vessels would have to be removed along with the cancer. If removing this tissue would mean leaving a limb that can't function well or would result in chronic pain, amputation may be the best option.

If the sarcoma has spread to distant sites (such as the lungs or other organs), all of the cancer will be removed if possible. That includes the original tumor plus the areas of spread. If it isn't possible to remove all of the sarcoma, then surgery may not be done at all.

Sometimes chemotherapy (chemo), radiation, or both is given before surgery. This, called *neoadjuvant* treatment, can shrink the tumor and allow it to be removed completely. Chemo or radiation can also be given before surgery to treat high-grade sarcomas when there is a great risk of the cancer spreading.

Most of the time, surgery cannot cure a sarcoma once it has spread. But if it has only spread to a few spots in the lung, the metastatic tumor can sometimes be removed. This can cure many patients, or at least lead to long-term survival.

You can read more about surgery for cancer in our document, *A Guide to Cancer Surgery*.

Radiation therapy for soft tissue sarcomas

Radiation therapy uses high-energy rays (such as x-rays) or particles to kill cancer cells.

Most of the time radiation is given after surgery as an added measure. This is called *adjuvant* treatment and it is done to kill any cancer cells that may be left behind after surgery.

Radiation may also be used before surgery to shrink the tumor and make the operation easier. This is called *neoadjuvant* treatment.

Radiation can be the main treatment for sarcoma in someone whose general health is too poor to undergo surgery.

Radiation therapy can also be used to help symptoms of sarcoma when it has spread. This is called *palliative treatment*.

Types of radiation therapy

External beam radiation therapy: For this treatment, radiation delivered from outside the body is focused on the cancer. This is the type of radiation therapy most often used to treat sarcomas. Treatments are often given daily, 5 days a week, usually for several weeks. Often a technique called *intensity modulated radiation therapy* (IMRT) is used. This better focuses the radiation on the cancer and lessens the impact on healthy tissue.

In some centers, proton beam radiation is an option. This uses streams of protons instead of x-ray beams to treat the cancer. Although this has some advantages over IMRT in theory, it hasn't been proven to be a better treatment for soft tissue sarcoma. Proton beam therapy is not widely available.

Intraoperative radiation therapy (IORT): This is a type of external beam radiation that is available in only a few centers. For this treatment, one large dose of radiation is given in the operating room after the tumor is removed but before the wound is closed. Giving radiation this way means that it doesn't have to travel through healthy tissue to get to the area that needs to be treated. It also allows nearby healthy areas to be shielded more easily from the radiation. Often, IORT is only one part of radiation therapy, and the patient receives some other type of radiation after surgery.

Brachytherapy: Brachytherapy (sometimes called *internal radiation therapy*) is a treatment that places small pellets (or seeds) of radioactive material in or near the cancer. For soft tissue sarcoma, these pellets are put into catheters (very thin tubes) that have been placed

during surgery. In high-dose rate (HDR) brachytherapy, the pellets give off a lot of radiation in a short time, and so stay in place for only minutes at a time. In low-dose rate (LDR) brachytherapy, the pellets may stay in place for days at a time, and are then removed.

Brachytherapy may be the only form of radiation therapy used or it can be combined with external beam radiation.

Side effects of radiation treatment

Side effects of radiation therapy depend on which area is treated and the dose given. Common side effects include

- Skin changes in the area the radiation went through the skin, which can range from redness to blistering and peeling
- Fatigue.
- Nausea and vomiting (more common with radiation to the abdomen)
- Diarrhea (most common with radiation to the pelvis and abdomen)
- Pain with swallowing (from radiation to the head, neck, or chest)
- Lung damage leading to problems breathing (from radiation to the chest)
- Bone weakness, which can lead to fracture years later

Radiation of large areas of an arm or leg can cause swelling, pain, and weakness in that limb.

Side effects of radiation therapy to the brain for metastatic sarcoma include hair loss, headaches, and problems thinking.

If given before surgery, radiation may cause problems with wound healing.

Many side effects improve or even go away some time after radiation is finished. Some though, like bone weakness and lung damage, can be permanent.

More information on this topic can be found in the Radiation section of our website, or in our document *Understanding Radiation Therapy: A Guide for Patients and Families*.

Chemotherapy for soft tissue sarcomas

Chemotherapy (chemo) is the use of drugs given into a vein or taken by mouth to treat cancer. These drugs enter the bloodstream and reach all areas of the body, making this treatment useful for cancer that has spread (metastasized) to other organs. Depending on the type and stage of sarcoma, chemotherapy may be given as the main treatment or as an adjuvant (addition) to surgery. Chemotherapy for soft tissue sarcoma generally uses a combination of several anti-cancer drugs.

The most commonly used drugs are ifosfamide (Ifex[®]) and doxorubicin (Adriamycin[®]). When ifosfamide is used, the drug mesna is also given. Mesna is not a chemo drug. It protects the bladder from the toxic effects of ifosfamide.

Other chemo drugs may be used as well, including cisplatin, dacarbazine (DTIC), docetaxel (Taxotere[®]), gemcitabine (Gemzar[®]), methotrexate, oxaliplatin, paclitaxel (Taxol[®]), vincristine, and vinorelbine (Navelbine[®]).

When several drugs are used together, the combination is given a shortened name such as: MAID (mesna, Adriamycin [doxorubicin], ifosfamide, and dacarbazine).

Chemotherapy drugs kill cancer cells but also damage some normal cells. Side effects depend on the type of drugs, the amount taken, and the length of treatment. Common chemo side effects include:

- Nausea and vomiting
- Loss of appetite
- Loss of hair
- Mouth sores
- Fatigue
- Low blood counts

Because chemotherapy can damage the blood-producing cells of the bone marrow, patients may have low blood cell counts. This can result in:

- Increased chance of infection (from too few white blood cells)
- Problems with bleeding or bruising (from too few blood platelets)
- Fatigue and weakness (from too few red blood cells)

Most side effects disappear once treatment is stopped. Hair will grow back after treatment ends, but it might look different. There are remedies for many of the temporary side effects of chemotherapy. For example, drugs can be given that prevent or reduce nausea and vomiting.

Some chemo side effects can last a long time or even be permanent. For example, doxorubicin can weaken the heart if too much is given. If you are to be treated with this drug, your doctor might check your heart function with special studies before starting this drug. The doctor will also watch the dose of doxorubicin closely during therapy.

Some chemo drugs cause nerve damage (called *neuropathy*), leading to numbness, tingling, or even pain in the hands and feet. To learn more about this see our document, *Peripheral Neuropathy After Chemotherapy*.

Chemotherapy may also permanently damage ovaries or testicles, which can lead to infertility (not being able to have children). This is discussed in more detail in our documents *Fertility and Women With Cancer* and *Fertility and Men With Cancer*.

Isolated limb perfusion

This procedure is a different way to give chemo. The circulation of the limb (arm or leg) with the tumor in it is separated from that of the rest of the body. Chemo is given just to that limb. Sometimes the blood is warmed up a bit to help the chemo work better (this is called *hyperthermia*). This can help shrink tumors, but it isn't clear that it helps patients live longer than standard chemo. It should only be done at centers with a lot of experience in giving chemo this way.

Targeted therapy for soft tissue sarcoma

Targeted therapy is a newer type of cancer treatment that uses drugs or other substances to identify and attack cancer cells while doing little damage to normal cells. These therapies attack the cancer cells' inner workings – the programming that makes them different from normal, healthy cells. Each type of targeted therapy works differently, but all alter the way a cancer cell grows, divides, repairs itself, or interacts with other cells.

Pazopanib (Votrient®)

At this time, pazopanib is the only targeted therapy drug approved to treat soft tissue sarcoma. It blocks several cellular enzymes called *tyrosine kinases* that are important for cell growth and survival. In a study of patients with advanced soft tissue sarcomas that had been treated with chemotherapy, pazopanib stopped the cancers from growing for an average of about 3 months longer than the patients given a sugar pill. So far, though, this drug hasn't been shown to help patients live longer. This drug is taken in pill form, once a day.

Common side effects include high blood pressure, nausea, diarrhea, headaches, low blood cell counts, and liver problems. In some patients this drug causes abnormal results on liver function tests, but it also rarely leads to severe liver damage that can be life threatening. Bleeding, clotting, and wound healing problems can occur, as well. This drug also rarely causes a problem with the heart rhythm or even a heart attack. If you are taking pazopanib, your doctor will monitor your heart with EKGs as well as check your blood tests to check for liver or other problems.

Imatinib (Gleevec®)

Imatinib is a tyrosine kinase inhibitor drug approved to treat gastrointestinal stromal tumors and some kinds of leukemia. It also can be helpful in treating desmoid tumors that can't be removed with surgery. Although it rarely causes tumors to shrink, it often causes them to stop growing for a time, which can be very helpful.

Side effects can include mild stomach upset, diarrhea, muscle pain, and skin rashes. The stomach upset is lessened if the drug is taken with food. Imatinib can also make people retain fluid. Often this causes some swelling in the face (around the eyes) or in the ankles. Rarely the drug causes more severe problems, such as fluid build up in the lungs or abdomen or causing problems with heart function.

Clinical trials for soft tissue sarcomas

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 website 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 website at www.cancer.gov/clinicaltrials.

You must meet the requirements to take part in any clinical trial. But if you do qualify for a clinical trial, you still decide whether or not to enter (enroll in) it.

Clinical trials are one way to get state-of-the-art cancer treatment. In some cases they may be the only way to get access to newer treatments. They are also 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 website or call our toll-free number (1-800-227-2345) and have it sent to you.

Complementary and alternative therapies for soft tissue sarcomas

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 social media groups and websites might 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 to 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 few or 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 that interest you. You can also read about them on the Complementary and Alternative Medicine page of our website.

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.

Treatment of soft tissue sarcomas, by stage

The only way to cure a soft tissue sarcoma is to remove it with surgery, so surgery is part of the treatment for all soft tissue sarcomas whenever possible. It is important that your surgeon and other doctors are experienced in the treatment of sarcomas. These are difficult tumors to treat and require both experience and expertise. Studies have shown that patients with sarcomas have better outcomes when they are treated at specialized cancer centers that have experience in sarcoma treatment.

Desmoid tumors

Desmoid tumors act differently from most soft-tissue sarcomas in that although they can grow into nearby tissues and often come back after surgery, they rarely spread to distant sites.

Some desmoid tumors can be watched without treatment for a time. Treatment will be given if the tumor is growing or is causing pain or other symptoms.

If treatment is needed and the entire tumor can be removed, the first treatment is often surgery. If the entire tumor is removed and the margins are clear, no other treatment is needed. These tumors can also be treated with radiation (instead of surgery).

For tumors that are large or have come back after treatment, drug therapy may be helpful. The drug sulindac, normally used to treat arthritis, can stop tumor growth or even cause the tumor to shrink. It can take months for the drug to work, but its effect can last for years. Drugs that block estrogen (tamoxifen and toremifene) have also been helpful in some patients. Some desmoid tumors have responded to treatment with chemotherapy (chemo) using the drug doxorubicin (Adriamycin), which may be used alone or with other drugs. The combination of methotrexate and vinblastine has also been helpful. Interferon, an immune-boosting drug, has also been used with some success. Another option is the targeted drug imatinib (Gleevec).

Stage I soft tissue sarcoma

Stage I soft tissue sarcomas are low-grade tumors of any size. Small (less than 5 cm or about 2 inches across) tumors of the arms or legs may be treated with surgery alone. The goal of surgery is to remove the tumor with some of the normal tissue around it. If cancer cells are found in or near the edges of the tissue removed (called *positive* or *close* margins), it can mean that some cancer was left behind. Often the best option for positive or close margins is more surgery. Another option is treating with radiation therapy after surgery. This lowers the chance of the cancer coming back.

If the tumor is not in a limb, (for example it is in the head, neck, or abdomen), removing the entire tumor with enough normal tissue around it can be more difficult. For these tumors, radiation with or without chemo may be given before surgery. This may be able to shrink the tumor enough to remove it entirely with surgery. If radiation is not used before surgery, it may be given after surgery to lessen the chance that the tumor will come back.

Stages II and III soft tissue sarcoma

Some stage III tumors have already spread to nearby lymph nodes. Most stage II and III sarcomas are high-grade tumors. They tend to grow and spread quickly. Even when these sarcomas have not yet spread to lymph nodes, the risk of spread (to lymph nodes or distant sites) is very high. These tumors also tend to grow back in the same area after they are removed (this is called *local recurrence*).

For all stage II and III sarcomas, surgically removing the tumor is still the main treatment. Lymph nodes will be removed as well if they contain cancer. If the tumor is large or in a place that would make surgery difficult, the patient may be treated with chemo, radiation, or both before surgery. For large tumors in the arms or legs, giving chemo by isolated limb perfusion is also an option. The goal of treatment is to shrink the tumor, making it easier to remove. These treatments also lower the chance of the tumor coming back in or near the same place it started. Smaller tumors may be treated with surgery first, then radiation to lower the risk of the tumor coming back. Sometimes chemo is given as well. When chemo is given, the drug most often used is doxorubicin (Adriamycin). This drug may be combined with ifosfamide (Ifex) and other drugs.

In rare cases, amputation is needed to remove the entire tumor. As with stage I sarcomas, radiation therapy with or without chemo can be used alone when the tumor's location or size or the patient's health in general makes surgery impossible. There is evidence that chemo after surgery may benefit some people with stage II and III sarcomas.

Stage IV soft tissue sarcoma

A sarcoma is considered stage IV when it has spread to distant sites (M1). Stage IV sarcomas are rarely curable. But some patients may be cured if the main tumor and all of the areas of cancer spread (metastases) can be removed by surgery. The best success rate is when it has spread only to the lungs. This is still an area where doctors disagree about which patients will benefit. Those patients' main tumors should be treated as in stages II or III, and metastases should be completely removed, if possible.

For patients whose primary tumor and all metastases cannot be completely removed by surgery, radiation therapy and/or chemotherapy are often given to relieve symptoms. The chemo drugs doxorubicin and ifosfamide are often the first choice — either alone or together with other drugs. Gemcitabine and docetaxel may be given if the first combination stops working (or doesn't work). Patients with angiosarcomas may benefit from treatment with paclitaxel (Taxol) or docetaxel (Taxotere) with vinorelbine (Navelbine).

Recurrent sarcoma

Cancer is called *recurrent* when it come backs after treatment. Recurrence can be local (in or near the same place it started) or distant (spread to other organs or tissues such as the lungs or brain). If the sarcoma comes back in the same area where it started, it may be treated with surgery. Radiation therapy is another option, especially if radiation wasn't part of the treatment of the original tumor. If external beam radiation was used before, brachytherapy may still be an option.

If the sarcoma returns at a distant site, chemo may be given. If the sarcoma has spread only to the lungs, it may be possible to remove all the areas of spread with surgery. Radiation is used to treat sarcomas that spread to the brain, as well as any recurrences that cause symptoms such as pain.

More treatment information for soft tissue sarcomas

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 information via telephone (1-800-4-CANCER) and its website (www.cancer.gov). Information for patients as well as more detailed information for cancer care professionals is also available on www.cancer.gov.

No cancer treatment can be done without consent. People who are capable of making their own decisions can decide whether they are willing to take any medical treatment, including any recommended cancer treatments. Your doctor should discuss treatment options with you, and explain what is likely to happen if you accept or refuse these treatments. For more, you can read our document *Informed Consent*.

What should you ask your doctor about soft tissue sarcomas?

As you cope with cancer and cancer treatment, you need to have honest, open discussions with your doctor. You should feel comfortable asking any question 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 sarcoma do I have?

- How much experience do you have in diagnosing and treating sarcoma?
- Has my cancer spread?
- What is the stage of my cancer and what does that mean?
- What are my treatment choices?
- What treatment do you recommend and why?
- What risks or side effects are there to the treatments you suggest?
- What are the chances my cancer will come back with these treatment plans?
- What should I do to be ready for treatment?
- What's my outlook?

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.

What happens after treatment for soft tissue sarcomas?

For some people with soft tissue sarcoma, 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 accept this uncertainty and are living 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. They might 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*, has 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 have and might do 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. This 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.

It is important to keep health insurance. Tests and doctor visits cost a lot, and even though no one wants to think about 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 to help you 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 that you 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 in the future. Make sure you have the following information handy:

- A copy of your pathology report(s) from any biopsies or surgery
- If you had surgery, a copy of your operative report(s)
- If you were hospitalized, a copy of the discharge summary that doctors prepare when a patient is sent home
- If you had radiation, a copy of your radiation treatment summary
- If you had drug treatment (such as chemotherapy hormone therapy, or targeted therapy), a list of your drugs, drug doses, and when you took them
- Copy of recent imaging studies (such as x-rays, CT scans, and MRI scans) on a DVD plus the radiology reports.

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

Lifestyle changes after having a soft tissue sarcoma

Having cancer and dealing with treatment can be time consuming and emotionally draining, but it can also 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 begin this process during cancer treatment.

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 might 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 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 might 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 to start healthy eating habits. 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. For more information, see our document, *Nutrition and Physical Activity During and After Cancer Treatment: Answers to Common Questions*.

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. 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. 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 a soft tissue sarcoma 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. You can also read more about the emotional issues common to people with cancer in our booklet, *Distress in People With Cancer*, which can be ordered from our toll-free number at no cost to you.

If treatment for soft tissue sarcoma 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 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. You can learn more about the changes that occur when curative treatment stops working, and about planning ahead for yourself and your family, in our documents called *Nearing the End of Life* and *Advance Directives*. You can read them online or call us to have free copies mailed to you.

At some point, you might 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 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 soft tissue sarcoma research and treatment?

Research is ongoing in the area of soft tissue sarcomas. Scientists are learning more about causes and ways to prevent sarcomas, and doctors are working to improve treatments.

Basic research

Scientists have made progress in understanding how certain changes in the DNA of soft tissue cells cause sarcomas to develop. This information is already being applied to new tests

to diagnose and classify sarcomas. This is important because accurate classification helps doctors select the most appropriate treatment. It is hoped that this information will soon lead to new strategies for treating these cancers, based on specific differences between normal and malignant soft tissue cells.

Classification

Classification of most cancers, including sarcomas, is based mostly on the way they look under a microscope. Recent research has shown that several different kinds of soft tissue sarcomas can look very similar under the microscope. By using new lab methods, researchers discovered that most cancers that used to be called *malignant fibrous histiocyoma* (MFH) are actually high-grade forms of liposarcoma, rhabdomyosarcoma, leiomyosarcoma, other sarcomas, and even carcinomas or lymphomas. About 10% to 15% of cancers called MFH before, still cannot be given a precise classification, and these are now called *pleomorphic undifferentiated sarcomas* or *undifferentiated pleomorphic sarcomas* (although the current classification system of the World Health Organization permits use of MFH as an alternate name).

Chemotherapy

Active research in chemotherapy for soft tissue sarcomas includes studies of new drugs and new ways to give drugs now available.

A new drug called *trabectedin* (Yondelis[®]) has been shown to help some patients with soft tissue sarcomas. It is approved for use in Europe, but it is still being tested in the United States. In this country, it is currently only available as part of a clinical trial.

Targeted therapy

Even more active than research into chemotherapy is research into targeted drugs. These drugs specifically block molecules in the cancer cells that cause the cancers to grow.

Other targeted drugs may also be helpful against sarcomas. For example, the targeted drug sunitinib (Sutent[®]) seems to slow the growth of many sarcomas. Sirolimus (Rapamune[®]) has shown some promise in treating patients with PEComa, especially pulmonary lymphangiomyomatosis. Other drugs being studied include cixutumumab, PD0332991, and ridaforolimus.

Anti-angiogenesis drugs

Drugs that block new blood vessel formation may help kill sarcomas by preventing their nourishment by the blood vessels. One such drug, bevacizumab (Avastin[®]) has shown a small benefit in sarcoma patients, when given with doxorubicin (Adriamycin). Cediranib is a

newer anti-angiogenesis drug that shows promise in treating a certain kind of soft tissue sarcoma.

Additional resources for soft tissue sarcoma

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 website, www.cancer.org.

Living with Cancer

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

Distress in People With Cancer

Talking With Friends and Relatives About Your Cancer (also in Spanish)

Helping Children When a Family Member Has Cancer: Dealing With Diagnosis (also available in Spanish)

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

Guide to Controlling Cancer Pain (also available in Spanish)

Genetic Testing: What You Need to Know

When Cancer Doesn't Go Away

Understanding cancer treatments

A Guide to Cancer Surgery (also available in Spanish)

A Guide to Chemotherapy (also available in Spanish)

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

Cancer treatment side effects

Fatigue in People With Cancer

Nausea and Vomiting

Anemia in People With Cancer

Peripheral Neuropathy Caused by Chemotherapy

Work, insurance, and finances

Health Insurance and Financial Assistance for the Cancer Patient

Returning to Work After Cancer Treatment

Working During Cancer Treatment

When cancer can't be cured, or if it comes back

When Your Cancer Comes Back: Cancer Recurrence

Advance Directives

Nearing the End of Life

Hospice Care

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 websites*

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

National Cancer Institute

Toll-free number: 1-800-4-CANCER or 1-800-422-6237; TTY: 1-800-332-8615

Website: www.cancer.gov

Offers current information about many types of cancer and treatments, as well as guidance for the family and children of people with cancer

The Sarcoma Alliance

Telephone number: 1-415-381-7236

Website: www.sarcomaalliance.org

Information about sarcoma, as well as a discussion board (for those who register non the website)

**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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