About Soft Tissue Sarcoma

Overview and Types

If you've been diagnosed with soft tissue sarcoma or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Is a Soft Tissue Sarcoma?

Research and Statistics

See the latest estimates for new cases of soft tissue sarcoma and deaths in the US and what research is currently being done.

- Key Statistics for Soft Tissue Sarcomas
- What's New in Soft Tissue Sarcoma Research?

What Is a Soft Tissue Sarcoma?

Cancer starts when cells start to grow out of control. Cells in nearly any part of the body can become cancer and can spread to other areas. To learn more about how cancers start and spread, see What Is Cancer?

There are many types of soft tissue tumors, and not all of them are cancerous. Many benign tumors are found in soft tissues. The word benign means they're not cancer. These tumors can't spread to other parts of the body. Some soft tissue tumors behave
in ways between a cancer and a non-cancer. These are called intermediate soft tissue tumors.

When the word sarcoma is part of the name of a disease, it means the tumor is malignant (cancer). A sarcoma is a type of cancer that starts in tissues like bone or muscle. Bone and soft tissue sarcomas are the main types of sarcoma. Soft tissue sarcomas can develop in 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 start 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 (belly) cavity (known as the retroperitoneum). Sarcomas are not common tumors.

Sarcomas that most often start in bones, such as osteosarcomas, and sarcomas that are most often seen in children, such as the Ewing Family of Tumors and Rhabdomyosarcoma, are not covered here.

Types of soft tissue sarcomas

There are more than 50 different types of soft tissue sarcomas. Some are quite rare, and not all are listed here:

- **Adult fibrosarcoma** usually affects fibrous tissue in the legs, arms, or trunk. It’s 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 start in legs.
- **Angiosarcoma** can start in blood vessels (hemangiosarcomas) or in 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 starts in tendons of the arms or legs. Under the microscope, it has some features of malignant melanoma, a type of cancer that starts in 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 teens and young adults. It’s found most often in the abdomen (belly).
- **Epithelioid sarcoma** most often starts in tissues under the skin of the hands, forearms, feet, or lower legs. Teens and young adults are often affected.
- **Fibromyxoid sarcoma, low-grade** is a slow-growing cancer that most often starts
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 sometimes called an **Evans' tumor**.

- **Gastrointestinal stromal tumor (GIST)** is a type of sarcoma that starts in the digestive tract. See [Gastrointestinal Stromal Tumor (GIST)](GIST) for more details.

- **Kaposi sarcoma** is a type of sarcoma that starts in the cells lining lymph or blood vessels. See [Kaposi Sarcoma](Kaposi Sarcoma).

- **Liposarcomas** are malignant tumors of fat tissue. They can start anywhere in the body, but they most often start in the thigh, behind the knee, and inside the back of the abdomen (belly). 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 start in 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.

- **Rhabdomyosarcoma** is the most common type of soft tissue sarcoma seen in children. See [Rhabdomyosarcoma](Rhabdomyosarcoma).

- **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** was once called **malignant fibrous histiocytoma** (MFH). It’s 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. It mostly tends to grow into other tissues around the place it started, but it can spread to distant parts of the body.

### Intermediate soft tissue tumors

These may grow and invade nearby tissues and organs, but they tend to not spread to other parts of the body.

- **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 fibromatosisor 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, like 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. 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 cancer (benign) but can be cancer (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 tumors, or tumors that are not cancer, can start in soft tissues. These include:

- **Elastofibromas**: benign tumors of fibrous tissue
- **Fibromas**: benign tumors of fibrous tissue
- **Fibrous histiocytomas**: benign tumors of fibrous tissue
- **Glomus tumors**: benign tumors that occur near blood vessels
- **Granular cell tumors**: usually benign tumors in adults that often start 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
- **Myxomas**: benign tumors that usually are in muscles but do 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 cancer. Neurofibromas are very common in people with an inherited condition called neurofibromatosis (also called von Recklinghausen disease) They’re much less common in people without this condition.
- **Neuromas**: benign tumors of nerves that can be painful
- **PEComas**: 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 lymphangioleiomyoma. Angiomyolipoma is a benign tumor that most often affects the kidney. Lymphangioleiomyomatosis (or LAM) is a rare disease of women in which the many lymphangioleiomyoma 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 tumors (also called nodular tenosynovitis)**: benign tumors of joint tissue

**Spindle cell tumors**

Spindle cell tumor and spindle cell sarcoma are descriptive names used because the cells look long and narrow 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 changes in soft tissues are caused by inflammation or injury and can form a mass that looks like a soft tissue tumor. Unlike a real tumor, they don’t come from a single abnormal cell, they have limited ability to grow or spread to nearby tissues, and never spread through the bloodstream or lymph system to other parts of the body. **Nodular**
fasciitis and myositis ossificans are 2 examples which affect tissues under the skin and muscle tissues, respectively.

Hyperlinks


References


Villalobos VM, Byfield SD, Ghate SR, Adejoro O. A retrospective cohort study of treatment patterns among patients with metastatic soft tissue sarcoma in the US. *Clin...*
Key Statistics for Soft Tissue Sarcomas

The American Cancer Society's estimates for soft tissue sarcomas in the United States for 2019 are (these statistics include both adults and children):

- About 12,750 new soft tissue sarcomas will be diagnosed (7,240 in males and 5,510 in females).
- About 5,270 Americans (2,840 males and 2,430 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
- Leiomyosarcoma

Certain types occur more often in certain parts of the body more often than others. For example, leiomyosarcomas are the most common type of sarcoma found in the abdomen (belly), while liposarcomas and undifferentiated pleomorphic sarcomas 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.

Visit the American Cancer Society's Cancer Statistics Center¹ for more key statistics.

Hyperlinks

1. [https://cancerstatisticscenter.cancer.org/](https://cancerstatisticscenter.cancer.org/)
What's New in Soft Tissue Sarcoma Research?

Research is ongoing in the area of soft tissue sarcomas. Because soft tissue sarcomas are rare and there are so many different types, it's been hard to study it well. Still, scientists are learning more about causes and genetic differences in types of sarcomas, and they're looking for ways to improve treatments.

Basic research

Scientists have made progress in understanding how certain gene changes in soft tissue cells cause sarcomas to develop. This information is already being used in new tests to diagnose and classify sarcomas. This is important because knowing the exact type of sarcoma will help doctors select treatment tailored for each person. It's hoped that this information will also lead to new ways to treat these cancers, based on specific differences between normal and cancer 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 a lot alike under the microscope. By using new tests, researchers have found that most cancers that used to be called malignant fibrous histiocytoma (MFH) are actually high-grade forms of liposarcoma, rhabdomyosarcoma\(^1\), leiomyosarcoma, other sarcomas, and even carcinomas or lymphomas. Tests to clearly classify the many types of soft tissue sarcoma are another key to deciding on the best treatment for each person.
Treatment

Researchers are looking for new and better ways to combine treatments, for example, using surgery, radiation, and chemo together, as well as new ways to treat soft tissue sarcomas.

Radiation

Doctors are looking at the best way to use radiation treatment. Studies are comparing radiation use before vs. after surgery to find out which has a greater impact on wound healing and long-term side effects. They're also looking at different types, doses, and schedules for radiation in an effort to find better and safer ways to use this treatment. Research is also being done to figure out when radiation is needed after surgery and when it's not.

Chemotherapy

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

Targeted therapy

There's a lot of active research on the use of targeted drugs. These drugs specifically block substances in or on cancer cells that cause the cancers to grow. Targeted treatments are used for many kinds of cancer and doctors are trying to find out if they might also be helpful against sarcomas.

Anti-angiogenesis drugs

Drugs that block new blood vessel formation may help kill sarcomas by keeping them from being fed by blood vessels. These drugs are being tested in many studies.

Other treatments

Many other treatments are being tested and are only available in clinical trials. Examples include vaccine treatments and T-cell therapies for people with advanced soft tissue sarcomas. The use of heat (hyperthermia) and cold (cryosurgery) to destroy tumors is also being explored. Most of these studies are in very early stages, and it will be awhile before doctors know that they work well enough to be part of regular treatment for soft tissue sarcoma.
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References


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Written by


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Soft Tissue Sarcoma Causes, Risk Factors, and Prevention

Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for soft tissue sarcoma.

- Risk Factors for Soft Tissue Sarcomas
- What Causes Soft Tissue Sarcomas?

Prevention

The only way to prevent some soft tissue sarcomas is to avoid exposure to risk factors whenever possible. Still, most sarcomas develop in people with no known risk factors. At this time, there's no known way to prevent this cancer. And for people getting radiation therapy, there's usually little choice.

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 many, doesn't mean that you will get the cancer. Also, many people get cancer without having a risk factor.
Injury and lifestyle factors such as smoking, diet, and exercise are NOT linked to the risk for soft tissue sarcoma. But the injury issue has caused some confusion in the past. One reason is that an injured area might swell. That swelling could look like a tumor, but it isn't one. Also, when you are injured, the pain may draw your attention to the injured area. A doctor may check the area, and x-rays or other imaging studies may be done. This can make it more likely that any sarcoma that's there will be discovered, even though it may have been there for some time.

Still, scientists have found a few risk factors that make a person more likely to develop soft tissue sarcomas:

**Radiation given to treat other cancers**

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

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 its risks. To learn more, see *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 is also known as *von Recklinghausen disease*. It usually runs in families and causes many benign (not cancer) tumors that form in nerves under the skin and in other parts of the body (These are called *neurofibromas*.) It’s caused by a defect (mutation) in genes called *NF1* and *NF2*. About 5% of people with
neurofibromatosis will develop a sarcoma 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 (FAP), 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 are discussed in *What Is a Soft Tissue Sarcoma?*²)

**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 cancer³, brain tumors⁴, leukemia⁵, and sarcomas. Still, only 10 to 20 out of 100 people with Li-Fraumeni syndrome will develop a soft tissue sarcoma. People with this syndrome are sensitive to the cancer-causing effects of radiation. So if they have a cancer that’s treated with radiation, they have a very high chance of developing a new cancer in the part of the body that was treated.

**Retinoblastoma**

*Retinoblastoma*⁶ is an eye cancer in children that can be caused by defects in the *RB1* gene. Children with this gene defect also have a higher risk of developing bone or soft tissue sarcomas, especially if the retinoblastoma was treated 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 is also called *nevoid basal cell carcinoma syndrome* (NBCCS). It’s caused by defects in the *PTCH1* gene. People with this syndrome have a high risk of developing many basal cell skin cancers⁷. They also have an increased risk of fibrosarcoma and rhabdomyosarcoma⁸.
Tuberous sclerosis

Tuberous sclerosis can be caused by a defect in the TSC1 and/or TSC2 gene. People with this syndrome often have seizures and learning problems. They get benign (not cancer) tumors in many different organs. They also have kidney problems, often along with a kidney tumor called angiomyolipoma. People with tuberous sclerosis have an increased risk of rhabdomyosarcoma.

Damaged lymph system

Lymph is a clear fluid containing immune system cells that’s 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 or damaged by radiation therapy, lymph fluid can build up and cause swelling. This is called lymphedema. Lymphangiosarcoma (a malignant (cancer) 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 hasn’t 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 isn’t known for certain. There’s no evidence that herbicides (weed killers) or insecticides, at levels encountered by the general public, cause sarcomas.

Hyperlinks

2. www.cancer.org/cancer/soft-tissue-sarcoma/about/soft-tissue-sarcoma.html

References


What Causes Soft Tissue Sarcomas?

Scientists don’t know exactly what causes most soft tissue sarcomas, but they have found some 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 genes in cells in the soft tissues.

Researchers have made great progress in understanding how certain changes in DNA (pieces of genes) can cause normal cells to become cancer. DNA carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. But DNA affects more than just the way we look.

The DNA is made of genes. Genes carry the recipes for making proteins, the molecules that control 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.

Many 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. Some of these syndromes are also linked to an increased risk of developing soft tissue sarcomas. (These syndromes were noted in Risk Factors for Soft Tissue Sarcomas) The syndromes are caused by defects (mutations) in genes that can be inherited (passed on) from a parent. Some of these gene defects can be found through testing. For more on this, see Family Cancer Syndromes¹ and Genetics and Cancer².

DNA mutations in soft tissue sarcoma are common. But they’re 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 don’t know why most soft tissue sarcomas develop in people who have no apparent risk factors.

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Can Soft Tissue Sarcomas Be Prevented?

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

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Written by


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Soft Tissue Sarcoma Early Detection, Diagnosis, and Staging

Detection and Diagnosis

Finding cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that's not always the case.

- Can Soft Tissue Sarcomas Be Found Early?
- Signs and Symptoms of Soft Tissue Sarcomas
- Tests for Soft Tissue Sarcomas

Stages and Outlook (Prognosis)

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and likely response to treatment.

- Soft Tissue Sarcoma Stages
- Survival Rates for Soft Tissue Sarcoma

Questions to Ask Your Cancer Care Team

Here are some questions you can ask your cancer care team to help you better understand your diagnosis and treatment options.

- Questions To Ask About Soft Tissue Sarcomas
Can Soft Tissue Sarcomas Be Found Early?

People who have a strong family history of soft tissue sarcomas or who have had other cancers when they were young, might want to talk to a doctor about the benefits and disadvantages of genetic testing\(^1\). 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 caused by defects in certain genes have an increased risk of soft tissue sarcomas. The changed (mutated) genes can be detected by genetic testing, so family members should discuss this option with their doctors. They should also see their doctor right away if they notice any lumps or growths. (The inherited conditions linked to soft tissue sarcoma are covered in Risk Factors for Soft Tissue Sarcomas\(^2\))

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 provider about any unexplained lumps or growths or other symptoms that may be caused by a soft tissue sarcoma.

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Signs and Symptoms of Soft Tissue Sarcomas

About half of soft tissue sarcomas start in an arm or leg. Most people notice a lump that's grown over time (weeks to months). The lump may or may not hurt.
When sarcomas grow in the back of the abdomen (the retroperitoneum), the symptoms often come from other problems the tumor is causing. For instance, they may cause blockage or bleeding of the stomach or bowels. They can press on nerves, blood vessels, or nearby organs. They can grow large enough for the tumor to be felt in the belly. Sometimes the tumors cause pain. About 4 of 10 sarcomas begin in the abdomen (belly).

In rare cases, sarcomas can start in the chest or in the head or neck.

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

- A new lump or a lump that's growing (anywhere on your body)
- Abdominal pain that's 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's digested, and it might 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.

References


Tests for Soft Tissue Sarcomas

If you have signs or symptoms that suggest you might have a soft tissue sarcoma, the doctor will likely need to do tests to find out if you have cancer.

Medical history and physical exam

The doctor will ask about your medical history, including your family history, to learn if you have any possible risk factors. You will also be asked about your symptoms, such as when they started and how long you’ve had them.

Imaging tests

Imaging tests use sound waves, x-rays, magnetic fields, or radioactive substances to create pictures of the inside of your body. Imaging tests may be done for a number of reasons, such as:

- To look at suspicious areas that might be cancer,
- To see if, and how far, cancer has spread
- To help determine if treatment is working

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.

**CT (computed tomography) scans**

A CT scan uses x-rays to make detailed cross-sectional images of your body. This test is often done if the doctor suspects a soft tissue sarcoma in the chest, abdomen (belly), or the retroperitoneum (the back of the abdomen). This test is also used to see if the sarcoma has spread to the lungs, liver, or other organs.

CT scans might be used to guide a biopsy needle into a tumor inside the body — the chest or abdomen, for example. This is called a **CT-guided needle biopsy**. (See below for more on biopsy.) You lie on the CT scanning table while a radiologist moves a biopsy needle toward the tumor. CT scans are repeated until the doctors are sure the needle is within the tumor.

**MRI (magnetic resonance imaging)**

MRI uses radio waves and strong magnets instead of x-rays to take pictures of the body. MRI scans are often part of the work-up of any tumor that could be a sarcoma. They’re often better than CT scans in evaluating sarcomas in the arms or legs.

MRI provides a good picture of the extent of the tumor. It can show your health care team many things about the tumor, like where it is, how big it is, and sometimes even the type of tissue it comes from (like bone, fat, or muscle). MRIs are also very helpful in examining the brain and spinal cord.

**Ultrasound**

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

Ultrasound may be done before a biopsy to see if a lump is a cyst, meaning if it has fluid in it and is likely not cancer, or if it’s solid and more likely a tumor. This test is often not needed if a CT or MRI was done.

**PET (positron emission tomography) scan**

PET scans use a form of radioactive sugar that’s put into the blood. Because cancers use glucose (sugar) at a higher rate than normal tissues, the radioactivity collects in the
cancer. A scanner can then spot the radioactive deposits.

A PET scan is useful when your doctor thinks the cancer has spread but doesn't know where. It can be used instead of many different x-rays because it scans your whole body. Often the PET scan is used with a CT scan (called a PET/CT scan). This helps decide if changes seen on the CT scan are cancer or something else. PET isn't often used for sarcoma, but it can be helpful in some cases.

**Biopsy**

If a soft tissue sarcoma is suspected based on exams and imaging tests, a biopsy is needed to know for sure that it's a sarcoma and not another type of cancer or a benign (not cancer) disease. In a biopsy, the doctor takes out a small piece of the tumor. This tissue is looked at under a microscope and other lab tests may be done as well.

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. See [Testing Biopsy and Cytology Specimens for Cancer](https://www.cancer.org/treatment/understanding-your-diagnosis/tests/testing-biopsy-and-cytology-specimens-for-cancer.html) to learn more about the types of biopsies, how the tissue is used in the lab to diagnose cancer, and what the results may show.

You might want to ask about your surgeon’s experience doing biopsies. 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.

**Hyperlinks**


**References**


Soft Tissue Sarcoma Stages

After someone is diagnosed with a soft tissue sarcoma, doctors will try to figure out if it has spread, and if so, how far. This process is called staging. The stage of a cancer describes how much cancer is in the body. It helps determine how serious the cancer is and how best to treat it. Doctors also use a cancer's stage when talking about survival statistics.

The stages of soft tissue sarcomas range from stages I (1) through IV (4). As a rule, the lower the number, the less the cancer has spread. A higher number, such as stage IV, means cancer has spread more. And within a stage, an earlier letter means a lower stage. Although each person's cancer experience is unique, cancers with similar stages tend to have a similar outlook and are often treated in much the same way.

How is the stage determined?

The staging system most often used for soft tissue sarcomas is the American Joint Committee on Cancer (AJCC) TNM system, which is based on 4 key pieces of information:

- The extent of the tumor (T): How large is the cancer?
• The spread to nearby lymph nodes (N): Has the cancer spread to nearby lymph nodes?
• The spread (metastasis) to distant sites (M): Has the cancer spread to distant organs such as the lungs?
• The grade (G) of the cancer: How much do the sarcoma cells look like normal cells?

Grade

The grade is partly used to determine the stage of a sarcoma. The staging system divides sarcomas into 3 grades (1 to 3). The grade of a sarcoma helps predict how rapidly it will grow and spread. It’s useful in predicting a patient’s outlook and helps determine treatment options.

The grade of a sarcoma is determined using a system known as the French or FNCLCC system, and is based on 3 factors:

• **Differentiation:** Cancer cells are given a score of 1 to 3, with 1 being assigned when they look a lot like 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).

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. Here’s what the grade numbers mean:

**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, 7 or 8

**Defining TNM**

There are different staging systems for soft tissue sarcomas depending on where the cancer is in the body.

- Head and neck
- Trunk and extremities (arms and legs)
- Abdomen and thoracic (chest) visceral organs
- Retroperitoneum

Numbers or letters after T, N, and M provide more details about each of these factors. Higher numbers mean the cancer is more advanced. Once a person’s T, N, and M categories have been determined, this information is combined in a process called stage grouping to assign an overall stage. Of the 4 main locations, only 2 (Trunk and Extremities and Retroperitoneum) have stage groupings. For more information see [Cancer Staging](#).

The staging system in the table below uses the pathologic stage (also called the surgical stage). It is determined by examining tissue removed during an operation. Sometimes, if surgery is not possible right away or at all, the cancer will be given a clinical stage instead. This is based on the results of a physical exam, biopsy, and imaging tests. The clinical stage will be used to help plan treatment. Sometimes, though, the cancer has spread further than the clinical stage estimates, and may not predict the patient’s outlook as accurately as a pathologic stage.

The system described below is the most recent AJCC system, effective January 2018. Cancer staging can be complex, so ask your doctor to explain it to you in a way you understand.

**Trunk and Extremities Sarcoma Stages**

<table>
<thead>
<tr>
<th>AJCC stage</th>
<th>Stage grouping</th>
<th>Trunk and Extremities Sarcoma Stage description</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>T1 N0 M0</td>
<td>The cancer is 5 cm (2 inches) or smaller (T1).</td>
</tr>
<tr>
<td></td>
<td></td>
<td>It has not spread to nearby lymph nodes (N0) or to distant sites (M0).</td>
</tr>
<tr>
<td>Stage</td>
<td>Tumor Size</td>
<td>Node Status</td>
</tr>
<tr>
<td>-------</td>
<td>------------</td>
<td>-------------</td>
</tr>
<tr>
<td>IB</td>
<td>T2, T3, T4</td>
<td>N0, M0</td>
</tr>
<tr>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>T1</td>
<td>N0, M0</td>
</tr>
<tr>
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<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IIIA</td>
<td>T2</td>
<td>N0, M0</td>
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<tr>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IIIB</td>
<td>T3 or T4</td>
<td>N0, M0</td>
</tr>
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</tbody>
</table>
The cancer is grade 2 (G2) or grade 3 (G3).

<table>
<thead>
<tr>
<th>IV</th>
<th>Any T N1 M0 Any G OR Any T Any N M1 Any G</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>The cancer is any size (Any T) AND it has spread to nearby lymph nodes (N1). It has not spread to distant sites (M0). It can be any grade.</td>
</tr>
<tr>
<td></td>
<td>The cancer is any size (Any T) AND it has spread to nearby lymph nodes (N1). It has spread to distant sites such as the lungs (M1). It can be any grade.</td>
</tr>
</tbody>
</table>

*The following categories are not listed in the table above:

- TX: Main tumor cannot be assessed due to lack of information.
- T0: No evidence of a primary tumor.
- NX: Regional lymph nodes cannot be assessed due to lack of information.

**Retroperitoneum Sarcoma Stages**

<table>
<thead>
<tr>
<th>AJCC stage</th>
<th>Stage grouping</th>
<th>Retroperitoneum Sarcoma Stage description*</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>T1 N0 M0 G1 or GX</td>
<td>The cancer is 5 cm (2 inches) or smaller (T1). It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 1 (G1) or the grade cannot be assessed (GX).</td>
</tr>
<tr>
<td>IB</td>
<td>T2, T3, T4</td>
<td>The cancer is: (T2, T3, T4)</td>
</tr>
</tbody>
</table>

11
<table>
<thead>
<tr>
<th>Stage</th>
<th>T</th>
<th>N</th>
<th>M</th>
<th>Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>IIA</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
<td>G2 or G3</td>
</tr>
<tr>
<td>IIIA</td>
<td>T3 or T4</td>
<td>N0</td>
<td>M0</td>
<td>G2 or G3</td>
</tr>
<tr>
<td>IIIB</td>
<td>Any T</td>
<td>N0</td>
<td>M0</td>
<td>G2 or G3</td>
</tr>
</tbody>
</table>

The cancer is:
- Larger than 5 cm but not more than 10 cm OR
- Larger than 10 cm but not more than 15 cm (T3) OR
- Larger than 15 cm (T4).

It has not spread to nearby lymph nodes (N0) or to distant sites (M0).

The cancer is grade 1 (G1) or the grade cannot be assessed (GX).

The cancer is grade 2 (G2) or grade 3 (G3).

The cancer is any size (Any T) AND it
**N1**

- M0
- Any G

has spread to nearby lymph nodes (N1).

It has not spread to distant sites (M0).

It can be any grade.

**IV**

- Any T
- Any N
- M1
- Any G

The cancer is any size (Any T) AND it has spread to nearby lymph nodes (N1).

It has spread to distant sites such as the lungs (M1). It can be any grade.

*The following categories are not listed in the table above:

- **TX:** Main tumor cannot be assessed due to lack of information.
- **T0:** No evidence of a primary tumor.
- **NX:** Regional lymph nodes cannot be assessed due to lack of information.

**Hyperlinks**

2. [www.cancer.org/treatment/understanding-your-diagnosis/staging.html](http://www.cancer.org/treatment/understanding-your-diagnosis/staging.html)

**References**


Last Medical Review: April 6, 2018 Last Revised: April 6, 2018
Survival Rates for Soft Tissue Sarcoma

Survival rates can give you an idea of what percentage of people with the same type and stage of cancer are still alive a certain amount of time (usually 5 years) after they were diagnosed. They can’t tell you how long you will live, but they may help give you a better understanding of how likely it is that your treatment will be successful.

Keep in mind that survival rates are estimates and are often based on previous outcomes of large numbers of people who had a specific cancer, but they can’t predict what will happen in any particular person’s case. These statistics can be confusing and may lead you to have more questions. Talk with your doctor about how these numbers may apply to you, as he or she is familiar with your situation.

What is a 5-year relative survival rate?

A relative survival rate compares people with the same type and stage of soft tissue sarcoma to people in the overall population. For example, if the 5-year relative survival rate for a specific stage of soft tissue sarcoma is 80%, it means that people who have that cancer are, on average, about 80% as likely as people who don’t have that cancer to live for at least 5 years after being diagnosed.

Where do these numbers come from?

The American Cancer Society relies on information from the SEER* database, maintained by the National Cancer Institute (NCI), to provide survival statistics for different types of cancer.

The SEER database tracks 5-year relative survival rates for soft tissue sarcoma in the United States, based on how far the cancer has spread. The SEER database, however, does not group cancers by AJCC TNM stages (stage 1, stage 2, stage 3, etc.). Instead, it groups cancers into localized, regional, and distant stages:

- **Localized**: The cancer is limited to the part of the body where it started.
- **Regional**: The cancer has spread to nearby structures or nearby lymph nodes.
- **Distant**: The cancer has spread to distant parts of the body such as the lungs.

5-year relative survival rates for soft tissue sarcoma
(Based on people diagnosed with soft tissue sarcoma between 2008 and 2014.)

<table>
<thead>
<tr>
<th>SEER Stage</th>
<th>5-Year Relative Survival Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Localized</td>
<td>81%</td>
</tr>
<tr>
<td>Regional</td>
<td>58%</td>
</tr>
<tr>
<td>Distant</td>
<td>16%</td>
</tr>
<tr>
<td>All SEER stages combined</td>
<td>65%</td>
</tr>
</tbody>
</table>

Understanding the numbers

- **These numbers apply only to the stage of the cancer when it is first diagnosed.** They do not apply later on if the cancer grows, spreads, or comes back after treatment.
- **These numbers don’t take everything into account.** Survival rates are grouped based on how far the cancer has spread, but your age, overall health, tumor grade, location where the tumor started (arm, leg, or retroperitoneum), how well the cancer responds to treatment, and other factors can also affect your outlook.
- **People now being diagnosed with soft tissue sarcoma may have a better outlook than these numbers show.** Treatments improve over time, and these numbers are based on people who were diagnosed and treated at least five years earlier.

*SEER= Surveillance, Epidemiology, and End Results

Hyperlinks


References

Questions To Ask 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 with 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?

Along with these examples, be sure to write down some of your own. For instance, you might want to know more about recovery times so that you can plan your work schedule. Or you may want to ask about second opinions or about clinical trials.

Hyperlinks

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Written by

The American Cancer Society medical and editorial content team (www.cancer.org/cancer/acs-medical-content-and-news-staff.html)

Our team is made up of doctors and oncology certified nurses with deep knowledge of cancer care as well as journalists, editors, and translators with extensive experience in medical writing.

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cancer.org | 1.800.227.2345
Treating Soft Tissue Sarcomas

If you’ve been diagnosed with a soft tissue sarcoma, your treatment team will discuss your options with you. It’s important to weigh the benefits of each treatment option against the possible risks and side effects.

How are soft tissue sarcomas treated?

The main types of treatment for soft tissue sarcoma are:

- Surgery for Soft Tissue Sarcomas
- Radiation Therapy for Soft Tissue Sarcomas
- Chemotherapy for Soft Tissue Sarcomas
- Targeted Therapy for Soft Tissue Sarcoma

Common treatment approaches

Treatment for a soft tissue sarcoma will depend on the type, location, and stage of the cancer, as well as your overall physical health. 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.

- Treatment of Soft Tissue Sarcomas, by Stage

Who treats soft tissue sarcomas?

Based on your treatment options, you might have different types of doctors on your treatment team. These doctors could include:

- An orthopedic surgeon: specializes in diseases of the bones, muscles, and joints
(for sarcomas in the arms and legs)

- **A surgical oncologist:** treats cancer with surgery (for sarcomas in the abdomen [belly] and retroperitoneum [the back of the abdomen])
- **A thoracic surgeon:** treats diseases of the lungs and chest with surgery (for sarcomas in the chest)
- **A medical oncologist:** treats cancer with medicines like chemotherapy
- **A radiation oncologist:** treats cancer with radiation therapy
- **A physiatrist (or rehabilitation doctor):** treats injuries or illnesses that affect how you move

You might have many other specialists on your treatment team as well, including physician assistants, nurse practitioners, nurses, nutrition specialists, social workers, and other health professionals.

- **Health Professionals Associated With Cancer Care**

**Making treatment decisions**

It’s important to discuss all treatment options, including their goals and possible side effects, with your doctors to help make the decision that best fits your needs. You may feel that you need to make a decision quickly, but it’s important to give yourself time to absorb the information you have learned. Ask your cancer care team questions.

If time permits, it is often a good idea to seek a second opinion. A second opinion can give you more information and help you feel more confident about the treatment plan you choose.

- **Questions To Ask About Soft Tissue Sarcomas**
- **Seeking a Second Opinion**

**Thinking about taking part in a clinical trial**

Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. 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 best way for doctors to learn better methods to treat cancer. Still, they’re not right for everyone.

If you would like to learn more about clinical trials that might be right for you, start by
asking your doctor if your clinic or hospital conducts clinical trials.

- **Clinical Trials**

### Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn’t mentioned to treat your cancer or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used along with your regular medical care. Alternative treatments are used instead of a doctor’s medical treatment. Although some of these methods might be helpful in relieving symptoms or helping you feel better, many have not been proven to work. Some might even be harmful.

Be sure to talk to your cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision.

- **Complementary and Alternative Medicine**

### Help getting through cancer treatment

Your cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services are an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help.

The American Cancer Society also has programs and services – including rides to treatment, lodging, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists.

- **Find Support Programs and Services in Your Area**

### Choosing to stop treatment or choosing no treatment at all

For some people, when treatments have been tried and are no longer controlling the cancer, it could be time to weigh the benefits and risks of continuing to try new
treatments. Whether or not you continue treatment, there are still things you can do to help maintain or improve your quality of life.

Some people, especially if the cancer is advanced, might not want to be treated at all. There are many reasons you might decide not to get cancer treatment, but it’s important to talk to your doctors and you make that decision. Remember that even if you choose not to treat the cancer, you can still get supportive care to help with pain or other symptoms.

• If Cancer Treatments Stop Working
• Palliative or Supportive Care

The treatment information given here is not official policy of the American Cancer 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.

Surgery for Soft Tissue Sarcomas

Surgery is commonly used to treat soft tissue sarcomas. Depending on the site and size of a sarcoma, surgery might be able to remove the cancer. 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 around it. 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 found at the edges of the removed tissue, it is said to have positive margins. This means that cancer cells may have been left behind. When cancer cells are left after surgery, more treatment such as radiation or another surgery -- might be needed.
• If cancer isn’t growing into the edges of the tissue removed, it's said to have negative or clear margins. The sarcoma has much less chance of coming back after surgery if it's removed with clear margins. In this case, surgery may be the only treatment needed.
When the tumor is in the abdomen, it can be hard to remove it and enough normal tissue to get clear margins because the tumor could be next to vital organs that can’t be taken out.

**Amputation and limb-sparing surgery**

In the past, many sarcomas in the arms and legs were treated by removing the limb (amputation). Today, this is rarely needed. Instead, the standard is surgery to remove the tumor without amputation. This is called **limb-sparing surgery**. A tissue graft or an implant may be used to replace the removed tissue. This might be followed by radiation therapy.

Sometimes, an amputation can’t be avoided. It might be the only way to remove all of the cancer. Other times, key 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 doesn’t work well or would result in chronic pain, amputation may be the best option.

**Surgery if sarcoma has spread**

If the sarcoma has spread to distant sites (like 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.

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

**Lymph node dissection**

If lymph nodes near the tumor are enlarged, cancer may be in them. During surgery, some of the swollen nodes may be sent to the lab and checked for cancer. If cancer is found, the lymph nodes in the area will be removed. Radiation might be used in that area after surgery.

**Treatments used with surgery**

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

Chemo and/or radiation may also be used **after** surgery. This is called **adjuvant** treatment. The goal is to kill any cancer cells that may be left in the body to lower the risk of the cancer coming back.

### More information about Surgery

For more general information about surgery as a treatment for cancer, see [Cancer Surgery](#).

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#).

### Hyperlinks

2. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html)

### References


MacNeill AJ, Gupta A, Swallow CJ. Randomized Controlled Trials in Soft Tissue


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**Radiation Therapy for Soft Tissue Sarcomas**

Radiation therapy uses high-energy rays (such as x-rays) or particles to kill cancer cells. It's a key part of soft tissue sarcoma treatment.

- Most of the time radiation is given after surgery. This is called **adjuvant treatment**. It's done to kill any cancer cells that may be left behind after surgery. Radiation can affect wound healing, so it may not be started until a month or so after surgery.
- Radiation may also be used before surgery to shrink the tumor and make it easier to remove. This is called **neoadjuvant** treatment.

Radiation can be the main treatment for sarcoma in someone who isn't healthy enough to have surgery. Radiation therapy can also be used to help ease symptoms of sarcoma when it has spread. This is called **palliative treatment**

**Types of radiation therapy**
• **External beam radiation**: 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. In most cases, a technique called **intensity modulated radiation therapy** (IMRT) is used. This better focuses the radiation on the cancer and lessens the damage to healthy tissue.

• **Proton beam radiation**: 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)**: 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 gets some other type of radiation after surgery.

• **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, soft tubes) that have been placed during surgery. 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 the part of the body treated and the dose given. Common side effects include:

• Skin changes where 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 belly)
• Diarrhea (most common with radiation to the pelvis and belly)
• 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 fractures or breaks 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 (in this case, it can be permanent), headaches, and problems thinking.

If given before surgery, radiation may cause problems with wound healing. If given after surgery, it can cause long-term stiffness and swelling that can affect how well the limb works.

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

**Chemoradiation**

After surgery, some high-grade sarcomas may be treated with radiation and chemotherapy at the same time. This is called **chemoradiation**.

This may also be done before surgery in cases where the sarcoma cannot be removed or removing it would cause major damage. Sometimes, chemoradiation can shrink the tumor enough to take care of these issues so it can be removed.

Chemoradiation can cause major side effects. And not all experts agree on its value in treating sarcoma. Radiation alone after surgery seems to works as well as chemoradiation. Still for some cases, this may be a treatment option to consider.

**More information about radiation therapy**

To learn more about how radiation is used to treat cancer, see [Radiation Therapy](#)².

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)³.

**Hyperlinks**

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. Different types of sarcoma respond better to chemo than others and also respond to different types of chemo. Chemotherapy for soft tissue sarcoma generally uses a combination of several anti-cancer drugs.

Chemo drugs used for sarcoma
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's used to protect the bladder from the toxic effects of ifosfamide.

Other chemo drugs may be used as well, including dacarbazine (DTIC), epirubicin, temozolomide (Temodal®), docetaxel (Taxotere®), gemcitabine (Gemzar®), vinorelbine (Navelbine®), trabectedin (Yondelis®), and eribulin (Halaven®).

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

**Isolated limb perfusion (ILP)**

ILP 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 then given just to that limb. Sometimes the blood is warmed up a bit to help the chemo work better (this is called hyperthermia). ILP may be used to treat tumors that cannot be removed or to treat high-grade tumors before surgery. It can help shrink tumors, but it isn’t clear that it helps patients live longer than standard chemo. ILP should only be done at centers with a lot of experience in giving chemo this way.

**Chemo side effects**

Chemotherapy drugs kill cancer cells but also damage some normal cells. This causes side effects. 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 go away over time once treatment is stopped. For instance, hair will grow back after treatment ends, but it might look different. There are treatments for many of the short-term side effects of chemo. For instance, 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 instance, doxorubicin can weaken the heart if too much is given. If you’re going to get this drug, your doctor might check your heart function with special studies before starting this drug. The doctor will also watch your heart function during therapy.

Some chemo drugs cause nerve damage (called neuropathy), leading to numbness, tingling, or even pain in the hands and feet. For more on this, see Peripheral Neuropathy Caused by Chemotherapy. 

Chemotherapy may also permanently damage ovaries or testicles, causing infertility (not being able to have children). This is covered in more detail in Fertility and Women With Cancer and Fertility and Men With Cancer.

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see Chemotherapy.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects.

Hyperlinks

1. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html


Targeted Therapy for Soft Tissue Sarcoma
Targeted therapy uses drugs or other substances to identify and attack sarcoma cells while doing little damage to normal cells. These therapies attack parts of cancer cells that make them different from normal, healthy cells. Each type of targeted therapy works differently, but all of them affect the way a cancer cell grows, divides, repairs itself, or interacts with other cells. Targeted therapy is an important part of treatment for many kinds of cancer. As doctors learn more about the biology of sarcoma cells, targeted therapy is becoming another treatment option for this cancer, too.

**Olaratumab (Lartruvo®)**

This drug is a type of monoclonal antibody, which is a man-made version of an immune system protein. It targets PDGFR-alpha, a protein on cancer cells that can help them grow. By blocking this protein, olaratumab can make some tumors shrink or stop growing. This has been shown to help people live longer.

This drug can be used along with the chemotherapy drug doxorubicin to treat soft tissue sarcomas that can’t be cured with radiation therapy or surgery.

Olaratumab is given by infusion into a vein (IV).

**Side effects**

Some people have allergic-like reactions while getting this drug, which can cause symptoms like low blood pressure, fever, chills, and rash. Less often, reactions can be more serious or even life-threatening.

Other possible side effects of this drug include nausea and vomiting, feeling tired, muscle or joint pain, swelling in the mouth or throat, hair loss, headache, loss of appetite, diarrhea, and nerve damage (neuropathy), which can cause numbness, tingling, or pain in the hands or feet.

**Pazopanib (Votrient®)**

Pazopanib blocks several cell enzymes called tyrosine kinases that are important for cell growth and survival. It may be used to treat certain advanced soft tissue sarcomas that have not responded to chemotherapy. It can help slow tumor growth and ease side effects in patients with sarcomas that cannot be removed with surgery. So far, though, it’s not clear that this drug helps patients live longer.

Pazopanib is taken in pill form, once a day.
Side effects

Common side effects include high blood pressure, fatigue, nausea, diarrhea, headaches, changes in hair color, low blood cell counts, and liver problems. In some patients this drug causes abnormal results on liver function tests, but it rarely leads to severe liver damage that can be life threatening.

Bleeding, clotting, and wound healing problems are rare, but can occur as well. This drug also rarely causes a problem with the heart rhythm or even a heart attack.

If you're taking pazopanib, your doctor will monitor your heart with EKGs and do blood tests to check for liver problems or other changes.

More information about targeted therapy

To learn more about how targeted drugs are used to treat cancer, see Targeted Cancer Therapy2.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects3.

Hyperlinks

2. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html

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Novartis Pharmaceuticals Corporation. Votrient (pazopanib): Highlights of Prescribing
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's important that your surgeon and other doctors are experienced in the treatment of sarcomas. These tumors are hard to treat and require both experience and expertise. Studies have shown that patients with sarcomas have better outcomes when they’re treated at specialized cancer centers that have experience in sarcoma treatment.

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's in the head, neck, or abdomen), it can be harder to take out the entire tumor with enough normal tissue around it. 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**

Most stage II and III sarcomas are high-grade tumors. They tend to grow and spread quickly. Some stage III tumors have already spread to nearby lymph nodes. 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’re removed. (This is called **local recurrence**.)

For all stage II and III sarcomas, surgically removing the tumor is the main treatment. Lymph nodes will also be removed if they contain cancer. Radiation may be given after surgery.

If the tumor is large or in a place that would make surgery difficult, but not in lymph nodes, 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. Chemo, radiation, or both might also be given after surgery. These treatments 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.

In rare cases, amputation is needed to remove the entire limb with the tumor.

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.

**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. Those patients’ main tumors should be treated as in stages II or III, and metastases should be completely removed, if possible. This is still an area where doctors disagree about what the best treatment is and which patients will benefit.
For patients whose primary tumor and all metastases cannot be completely removed by surgery, radiation therapy and/or chemotherapy are often used to relieve symptoms. The chemo drugs doxorubicin and ifosfamide are often the first choice — either together or along with other drugs. If doxorubicin is used, it might be given along with the targeted drug olaratumab (Lartruvo). 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 comes back 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 may be given after surgery, 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.

**References**


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After Soft Tissue Sarcoma Treatment

Living as a Cancer Survivor

For many people, cancer treatment often raises questions about next steps as a survivor.

- Living As a Soft Tissue Sarcoma Survivor

Cancer Concerns After Treatment

Treatment may remove or destroy the cancer, but it's very common to have questions about cancer coming back or treatment no longer working.

- Second Cancers After Soft Tissue Sarcoma

Living As a Soft Tissue Sarcoma Survivor

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. This is a very common if you've had cancer.

For other people, the cancer may never go away completely. They might get regular
treatments\textsuperscript{1} with chemotherapy, radiation therapy, or other therapies to try to help keep the cancer in check. Learning to live with cancer\textsuperscript{2} that does not go away can be difficult and very stressful.

Life after cancer means returning to some familiar things and also making some new choices.

Follow-up care

When treatment ends, your doctors will still want to watch you closely. It's 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\textsuperscript{3} 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.

At first these visits may be every 3 to 6 months. After 2 to 3 years, you may go to an every 6 month schedule for another few years. You can expect at least yearly check-ups for a long time after that. Chest x-rays and other imaging tests\textsuperscript{4} of the place the tumor was will be done at some of these visits. This helps the doctor watch for any signs that the sarcoma has come back.

During this time, it's very important to report any new symptoms to the doctor right away so that any problems can be found early, when they're easier to treat.

Depending on the type of treatment you had, physical therapy and rehabilitation may be a very important part of recovery.

Ask your doctor for a survivorship care plan

Talk with your doctor about developing a survivorship care plan\textsuperscript{5} for you. This plan might include:

- A suggested schedule for follow-up exams and tests
- A schedule for other tests you might need in the future, such as early detection (screening) tests for other types of cancer, or tests to look for long-term health effects from your cancer or its treatment
- A list of possible late- or long-term side effects from your treatment, including what
to watch for and when you should contact your doctor
• Diet and physical activity suggestions
• Reminders to keep your appointments with your primary care provider (PCP), who will monitor your general health care

Keeping health insurance and copies of your medical records

Even after treatment, it’s very important to keep health insurance. Tests and doctor visits cost a lot, and even though no one wants to think of their cancer coming back, this could happen.

At some point after your cancer treatment, you might see a new doctor who doesn’t know your medical history. It’s important to keep copies of your medical records to be able to give your new doctor the details of your diagnosis and treatment. Learn more in Keeping Copies of Important Medical Records.

Can I lower my risk of soft tissue sarcoma progressing or coming back?

If you have (or have had) a soft tissue sarcoma, you probably want to know if there are things you can do that might lower your risk of the cancer growing or coming back, such as exercising, eating a certain type of diet, or taking nutritional supplements. Unfortunately, it’s not yet clear if there are things you can do that will help.

Adopting healthy behaviors such as not smoking, eating well, getting regular physical activity, and staying at a healthy weight might help, but no one knows for sure. However, we do know that these types of changes can have positive effects on your health that can extend beyond your risk of soft tissue sarcoma or other cancers.

About dietary supplements

So far, no dietary supplements (including vitamins, minerals, and herbal products) have been shown to clearly help lower the risk of soft tissue sarcoma progressing or coming back. This doesn’t mean that no supplements will help, but it’s important to know that none have been proven to do so.

Dietary supplements are not regulated like medicines in the United States – they do not have to be proven effective (or even safe) before being sold, although there are limits on what they’re allowed to claim they can do. If you’re thinking about taking any type of
nutritional supplement, talk to your health care team. They can help you decide which ones you can use safely while avoiding those that might be harmful.

**If the cancer comes back**

If the cancer does recur at some point, your treatment options will depend on where the cancer is located, what treatments you’ve had before, and your health. For more information on how recurrent cancer is treated, see Treatment of Soft Tissue Sarcomas, by Stage.

For more general information on recurrence, you may also want to see [Understanding Recurrence](#).

**Could I get a second cancer after treatment?**

People who’ve had a soft tissue sarcoma can still get other cancers. In fact, sarcoma survivors are at higher risk for getting some other types of cancer. Learn more in [Second Cancers After Soft Tissue Sarcoma](#).

**Getting emotional support**

Some amount of feeling depressed, anxious, or worried is normal when cancer is a part of your life. Some people are affected more than others. But everyone can benefit from help and support from other people, whether friends and family, religious groups, support groups, professional counselors, or others. Learn more in [Life After Cancer](#).

**Hyperlinks**

3. [www.cancer.org/treatment/understanding-your-diagnosis/tests.html](http://www.cancer.org/treatment/understanding-your-diagnosis/tests.html)
Second Cancers After Soft Tissue Sarcoma

Cancer survivors can be affected by a number of health problems, but often their greatest concern is facing cancer again. If a cancer comes back after treatment it is called a *recurrence*. But some cancer survivors may develop a new, unrelated cancer later. This is called a *second cancer*. No matter what type of cancer you have had, it's still possible to get another (new) cancer, even after surviving the first.

Unfortunately, being treated for cancer doesn’t mean you can’t get another cancer. People who have had cancer can still get the same types of cancers that other people get. In fact, certain types of cancer and cancer treatments can be linked to a higher risk of certain second cancers.
Survivors of soft tissue sarcoma can get any type of second cancer, but they have an increased risk of these cancers, depending on where the tumor was and the type of treatment used:

- A second soft-tissue sarcoma (this is different than the first one coming back)
- Bone cancer
- Stomach cancer
- Thyroid cancer
- Melanoma of the skin
- Acute myeloid leukemia (AML)

Some second bone cancers may be due to treatment with radiation therapy. Radiation and chemotherapy likely contribute to the cases of leukemia.

**What you can do**

After treatment for soft tissue sarcoma, you should still see your doctor regularly. Tests will be done to look for signs the cancer has come back or spread. But experts do not recommend any additional testing to look for second cancers in patients without symptoms. Let your doctor know about any new symptoms or problems, because they could be caused by the cancer coming back, by a new disease, or by a second cancer.

Patients who have completed treatment should follow the American Cancer Society guidelines for the early detection of cancer.

The Children’s Oncology Group has guidelines for the follow-up of patients treated for cancer as a child, teen, or young adult, including screening for second cancers. These can be found at [www.survivorshipguidelines.org](http://www.survivorshipguidelines.org).

All survivors of soft tissue sarcoma should avoid tobacco smoke, as smoking increases the risk of many cancers, as well as other health problems.

To help maintain good health, survivors should also:

- Get to and stay at a healthy weight
- Adopt a physically active lifestyle
- Consume a healthy diet, with an emphasis on plant foods
- Limit alcohol use to no more than 1 drink per day for women or 2 per day for men
These steps may also lower the risk of some cancers.

See Second Cancers in Adults\textsuperscript{12} for more information.

**Hyperlinks**


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**Written by**


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