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# 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?](#)
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# 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?](#)<sup>1</sup>

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](#)<sup>2</sup>, and sarcomas that are most often seen in children, such as the [Ewing Family of Tumors](#)<sup>3</sup> and [Rhabdomyosarcoma](#)<sup>4</sup>, 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](#)<sup>5</sup>.
- **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](#)<sup>6</sup>, 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 \(GIST\)](#)<sup>7</sup> for more details.
- **Kaposi sarcoma** is a type of sarcoma that starts in the cells lining lymph or blood vessels. See [Kaposi Sarcoma](#)<sup>8</sup>.
- **Leiomyosarcoma** is a type of cancer that starts in smooth muscle tissue. These tumors often start in the abdomen, but they can also start in other parts of the body, such as the arms or legs, or in the uterus (see [Uterine Sarcoma](#)<sup>9</sup>).
- **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](#)<sup>10</sup>.
- **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 (UPS)** 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 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, 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 **lymphangioliomyoma**. Angiomyolipoma is a benign tumor that most often affects the kidney. Lymphangioliomyomatosis (or LAM) is a rare disease of women in which the many lymphangioliomyoma 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

1. [www.cancer.org/cancer/cancer-basics/what-is-cancer.html](http://www.cancer.org/cancer/cancer-basics/what-is-cancer.html)
2. [www.cancer.org/cancer/osteosarcoma.html](http://www.cancer.org/cancer/osteosarcoma.html)
3. [www.cancer.org/cancer/ewing-tumor.html](http://www.cancer.org/cancer/ewing-tumor.html)
4. [www.cancer.org/cancer/rhabdomyosarcoma.html](http://www.cancer.org/cancer/rhabdomyosarcoma.html)
5. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/lymphedema.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/lymphedema.html)
6. [www.cancer.org/cancer/melanoma-skin-cancer.html](http://www.cancer.org/cancer/melanoma-skin-cancer.html)
7. [www.cancer.org/cancer/gastrointestinal-stromal-tumor.html](http://www.cancer.org/cancer/gastrointestinal-stromal-tumor.html)
8. [www.cancer.org/cancer/kaposi-sarcoma.html](http://www.cancer.org/cancer/kaposi-sarcoma.html)
9. [www.cancer.org/cancer/uterine-sarcoma.html](http://www.cancer.org/cancer/uterine-sarcoma.html)
10. [www.cancer.org/cancer/rhabdomyosarcoma.html](http://www.cancer.org/cancer/rhabdomyosarcoma.html)

## References

Helman LJ, Maki RG. Sarcomas of soft tissue. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, PA. Elsevier: 2014: 1753-1791.

MacNeill AJ, Gupta A, Swallow CJ. Randomized Controlled Trials in Soft Tissue Sarcoma: We Are Getting There! *Surg Oncol Clin Am*. 2017;26:531-544.

Medscape. Benign and Malignant Soft-Tissue Tumors. March 27, 2017. Accessed at <https://emedicine.medscape.com/article/1253816-overview> on March 27, 2018.

National Comprehensive Cancer Network, Clinical Practice Guidelines in Oncology (NCCN Guidelines®), Soft Tissue Sarcoma, Version 1.2018 -- October 31, 2017. Accessed at [www.nccn.org/professionals/physician\\_gls/pdf/sarcoma.pdf](http://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf) on March 27, 2018.

Singer S, Maki R, O'Sullivan B. Soft tissue sarcoma In: DeVita VT, Heilman S, Rosenberg SA, eds. *Cancer: Principles and Practice of Oncology*. 9th ed. Philadelphia,

Pa: Lippincott Williams & Wilkins; 2011:1533-1577.

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 Sarcoma Res.* 2017;7:18.

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## Key Statistics for Soft Tissue Sarcomas

The American Cancer Society's estimates for soft tissue sarcomas in the United States for 2020 are:

- About 13,130 new soft tissue sarcomas will be diagnosed (7,470 in males and 5,660 in females).
- About 5,350 people (2,870 males and 2,480 females) are expected to die of soft tissue sarcomas.

These statistics include both adults and children.

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 Center](#)<sup>1</sup> for more key statistics.

## Hyperlinks

1. <https://cancerstatisticscenter.cancer.org/>

## References

American Cancer Society. *Cancer Facts & Figures 2020*. Atlanta, Ga: American Cancer Society; 2020.

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# 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 has 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](#)<sup>1</sup>, 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.

## Hyperlinks

1. [www.cancer.org/cancer/rhabdomyosarcoma.html](http://www.cancer.org/cancer/rhabdomyosarcoma.html)

## References

Fiore M, Ford S, Callegaro D, et al. Adequate Local Control in High-Risk Soft Tissue Sarcoma of the Extremity Treated with Surgery Alone at a Reference Centre: Should Radiotherapy Still be a Standard? *Ann Surg Oncol*. 2018 Feb 22.

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## Written by

The American Cancer Society medical and editorial content team  
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