



# About Soft Tissue Sarcoma

## Overview and Types

If you have 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 and Treatment?](#)

## What Is a Soft Tissue Sarcoma?

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

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

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

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

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

- Adult fibrosarcoma
- Alveolar soft-part sarcoma
- Angiosarcoma (includes hemangiosarcoma and lymphangiosarcoma)
- Clear cell sarcoma
- Desmoplastic small round cell tumor
- Epithelioid sarcoma
- Fibromyxoid sarcoma, low-grade
- Gastrointestinal stromal tumor (GIST): This is a type of sarcoma that develops in the digestive tract. It is covered in [Gastrointestinal Stromal Tumor \(GIST\)](#).
- Kaposi sarcoma: This is a type of sarcoma that develops from the cells lining lymph or blood vessels. It is covered in [Kaposi Sarcoma](#).
- Liposarcoma (includes dedifferentiated, myxoid, and pleomorphic liposarcomas)
- Leiomyosarcoma
- Malignant mesenchymoma
- Malignant peripheral nerve sheath tumors (includes neurofibrosarcomas, neurogenic sarcomas, and malignant schwannomas)
- Myxofibrosarcoma, low-grade
- Rhabdomyosarcoma: This is the most common type of soft tissue sarcoma seen in children and is discussed in [Rhabdomyosarcoma](#).
- Synovial sarcoma
- Undifferentiated pleomorphic sarcoma (previously known as malignant fibrous histiocytoma or MFH)

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

Intermediate soft tissue tumors include:

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

## Types of soft tissue sarcomas

- **Adult fibrosarcoma** usually affects fibrous tissue in the legs, arms, or trunk. It is most common in people between the ages of 20 and 60, but can occur in people of any age, even in infants.
- **Alveolar soft-part sarcoma** is a rare cancer that mostly affects young adults. These tumors most commonly occur in legs.
- **Angiosarcoma** can develop either from blood vessels (*hemangiosarcomas*) or from lymph vessels (*lymphangiosarcomas*). These tumors sometimes start in a part of the body that has been treated with radiation. Angiosarcomas are sometimes seen in the breast after radiation therapy and in limbs with [lymphedema](#).
- **Clear cell sarcoma** is a rare cancer that often develops in tendons of the arms or legs. Under the microscope, it has some features of [malignant melanoma](#), a type of cancer that develops from pigment-producing skin cells. How cancers with these features start in parts of the body other than the skin is not known.
- **Desmoplastic small round cell tumor** is a rare sarcoma of adolescents and young adults, found most often in the abdomen.
- **Epithelioid sarcoma** most often develops in tissues under the skin of the hands, forearms, feet, or lower legs. Adolescents and young adults are often affected.
- **Fibromyxoid sarcoma, low-grade** is a slow growing cancer that most often develops as a painless growth in the trunk or arms and legs (particularly the thigh). It is more common in young to middle aged adults. It is also sometimes called an *Evans' tumor*.
- **Liposarcomas** are malignant tumors of fat tissue. They can develop anywhere in the body, but they most often develop in the thigh, behind the knee, and inside the back of the abdomen. They occur mostly in adults between 50 and 65 years old.
- **Malignant mesenchymoma** is a rare type of sarcoma that shows features of fibrosarcoma and features of at least 2 other types of sarcoma.
- **Malignant peripheral nerve sheath tumors** include neurofibrosarcomas, malignant schwannomas, and neurogenic sarcomas. These are sarcomas that

develop from the cells that surround a nerve.

- **Myxofibrosarcomas, low-grade** are most often found in the arms and legs of elderly patients. They are most common in or just under the skin and there might be more than one tumor nodule.
- **Synovial sarcoma** is a malignant tumor of the tissue around joints. The most common locations are the hip, knee, ankle, and shoulder. This tumor is more common in children and young adults, but it can occur in older people.
- **Undifferentiated pleomorphic sarcoma**, previously known as *malignant fibrous histiocytoma* (MFH), is most often found in the arms or legs. Less often, it can start inside at the back of the abdomen (the retroperitoneum). This sarcoma is most common in older adults. Although it mostly tends to grow locally, it can spread to distant sites.

## Intermediate soft tissue tumors

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

tissue surrounding the lung (called the pleura). Many tumors that were once called hemangiopericytomas are now considered solitary fibrous tumors.

## Benign soft tissue tumors

Many benign (non-cancerous) tumors can occur in soft tissues as well. 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 occur often in the tongue but can be found almost anywhere in the body.
- Hemangiomas: benign tumors of blood vessels
- Hibernomas: benign tumors of fat tissue
- Lipomas: very common benign tumors of fat tissue
- Leiomyomas: benign tumors of smooth muscle that can be found anywhere in the body but are very common in the walls of the uterus where they are known as *fibroids*.
- Lipoblastomas: benign fat tissue tumors most often seen in children
- Lymphangiomas: benign tumors of lymph vessels
- Myxomas: benign tumors that usually are located 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 cancerous. Neurofibromas are very common in people with an inherited condition called neurofibromatosis (also called von Recklinghausen disease), and are much less common in people without this condition.
- Neuromas: benign 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 lymphangiomyoma. Angiomyolipoma is a benign tumor that most often affects the kidney. Lymphangiomyomatosis (LAM) is a rare disease of women in which the many lymphangiomyoma tumors grow into the lung tissue and interfere with lung function.
- Rhabdomyomas: benign tumors of skeletal and heart muscle

- Schwannomas (neurilemmomas): benign tumors of the cells that coat nerves
- Tenosynovial giant cell tumors (also called nodular tenosynovitis): benign tumors of joint tissue

## Spindle cell tumors

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

## Tumor-like conditions of soft tissue

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

- [References](#)

[See all references for Soft Tissue Sarcoma](#)

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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 2018 are (these statistics include both adults and children):

- About 13,040 new soft tissue sarcomas will be diagnosed (7,370 cases in males and 5,670 cases in females).

- 5,150 Americans (2,770 males and 2,380 females) are expected to die of soft tissue sarcomas.

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

Visit the American Cancer Society's [Cancer Statistics Center](#) for more key statistics.

- [References](#)

[See all references for Soft Tissue Sarcoma](#)

American Cancer Society. *Cancer Facts & Figures 2018*. Atlanta, Ga: American Cancer Society; 2018.

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## What's New in Soft Tissue Sarcoma Research and Treatment?

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

### Basic research

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

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

## Classification

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

## Chemotherapy

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

For example, a new drug called *trabectedin* (Yondelis<sup>®</sup>) has been shown to help some patients with certain types of soft tissue sarcomas, and has recently been approved for use in the United States.

## Targeted therapy

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

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



## Anti-angiogenesis drugs

Drugs that block new blood vessel formation may help kill sarcomas by preventing their nourishment by the blood vessels. One such drug, bevacizumab (Avastin<sup>®</sup>) has shown a small benefit in sarcoma patients, when given with doxorubicin (Adriamycin). Cediranib is a newer anti-angiogenesis drug that shows promise in treating a certain kind of soft tissue sarcoma.

- [References](#)

[See all references for Soft Tissue Sarcoma](#)

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