

# **About Soft Tissue Sarcoma**

Get an overview of soft tissue sarcoma in adults and the latest key statistics in the US.

#### **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?

- Types of soft tissue sarcomas
- Intermediate soft tissue tumors
- Benign soft tissue tumors
- Spindle cell tumors

• Tumor-like conditions of soft tissue

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 <u>What Is Cancer?</u><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.

Sarcomas that most often start in bones, such as <u>osteosarcomas</u><sup>2</sup>, and sarcomas that are most often seen in children, such as the <u>Ewing family of tumors</u><sup>3</sup> and <u>rhabdomyosarcomas</u><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 the 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.

### Intermediate soft tissue tumors

These tumors 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 tumor 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 tumors with features in between fibrosarcoma and benign tumors such as fibromas (see below). They tend to grow slowly but often steadily. They are also called **desmoid tumors**, as well as by the more scientific name musculoaponeurotic fibromatosis,or just aggressive fibromatosis. They rarely, if ever, spread to distant parts of the body, but they can cause problems by growing into nearby tissues. They can sometimes even be fatal. Some doctors consider them a type of low-grade fibrosarcoma, but others believe they are a unique type of fibrous tissue tumor. Certain hormones, like estrogen, make some desmoid tumors grow. Anti-estrogen drugs are sometimes useful in treating desmoids that cannot be removed completely 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 benign (not cancer), but some are cancer (malignant). Some start in the thigh, underarm, and pelvis. They can also start in the tissue lining the lungs (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 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:

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 they never spread to other parts of the body. **Nodular fasciitis** and **myositis ossificans**are 2 examples. They affect tissues under the skin and muscle tissues, respectively.

# **Hyperlinks**

(NCCN Guidelines®), Soft Tissue Sarcoma, Version 1.2018 -- October 31, 2017. Accessed at 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.

Last Revised: November 23, 2021

# **Key Statistics for Soft Tissue Sarcomas**

The American Cancer Society's estimates for soft tissue sarcomas in the United States for 2024 are:

- About 13,590 new soft tissue sarcomas will be diagnosed (7,700 in males and 5,890 in females).
- About 5,200 people (2,760 males and 2,440 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 <u>Cancer Statistics Center<sup>1</sup></u> for more key statistics.

# **Hyperlinks**

1. cancerstatisticscenter.cancer.org/

#### References

American Cancer Society. *Cancer Facts & Figures 2024*. Atlanta: American Cancer Society; 2024.

Last Revised: January 12, 2023

# What's New in Soft Tissue Sarcoma Research?

- Basic research
- Classification
- Treatment

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**

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/types/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.

Last Revised: April 6, 2018

#### Written by

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

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.

\_\_\_\_\_