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 Drug Therapy for Soft Tissue Sarcoma
- Immunotherapy 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:

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 to relieve symptoms or treat your cancer that your doctors haven't mentioned. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

helpline specialists. Or, if you prefer, you can use our chat feature on cancer.org to connect with one of our specialists.

- Palliative Care
- Programs & Services

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 as 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

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 your cancer care team any questions you may have about your treatment options.

Surgery for Soft Tissue Sarcomas

- Amputation and limb-sparing surgery
- Surgery if sarcoma has spread
- Treatments used with surgery
- More information about Surgery

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

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

Management of Resectable Soft Tissue Sarcoma. *Ann Surg Oncol*. 2017;24(2):425-433.

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.

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.

Keung EZ, Chiang YJ, Voss RK, et al. Defining the incidence and clinical significance of lymph node metastasis in soft tissue sarcoma. *Eur J Surg Oncol*. 2018;44(1):170-177.

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.

National Cancer Institute. Adult Soft Tissue Sarcoma Treatment (PDQ®)–Patient Version. May 15, 2017. Accessed at www.cancer.gov/types/soft-tissue-sarcoma/patient/adult-soft-tissue-treatment-pdq on March 30, 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 on March 30, 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.

Last Revised: April 6, 2018

Radiation Therapy for Soft Tissue Sarcomas

- Types of radiation therapy
- Side effects of radiation treatment

- Chemoradiation
- More information about radiation therapy

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 treatmen**t. 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[1].

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 **intensisaGj 0/lated radiation therapy : This is th7P572tment**

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

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

1. www.cancer.org/cancer/managing-cancer/palliative-care.html
2. www.cancer.org/cancer/managing-cancer/treatment-types/radiation.html
3. www.cancer.org/cancer/managing-cancer/side-effects.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.

Lehane C, Ho F, Thompson SR, et al. Neoadjuvant chemoradiation (modified Eilber protocol) versus adjuvant radiotherapy in the treatment of extremity soft tissue sarcoma. *J Med Imaging Radiat Oncol*. 2016;60(4):539-544.

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 on March 30, 2018.

Nesseler JP, Salleron J, Rios M, et al. A retrospective cohort study to assess adjuvant concurrent chemoradiation (CCRT) compared to adjuvant radiation therapy (RT) in the treatment of grade 2 and 3 extremity soft tissue sarcomas. *Radiother Oncol*. 2017;125(1):160-167.

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.

Last Revised: April 6, 2018

Chemotherapy for Soft Tissue Sarcomas

- Chemo drugs used for sarcoma
- Chemo side effects
- More information about chemotherapy

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 (Temodar®), docetaxel (Taxotere®), gemcitabine (Gemzar®), vinorelbine (Nav

Neuropathy Caused by Chemotherapy[4].

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[5] and Fertility and Men With Cancer[6].

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see Chemotherapy[7].

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects[8].

Hyperlinks

1. www.cancer.org/cancer/managing-cancer/side-effects.html
2. www.cancer.org/cancer/managing-cancer/side-effects/infections.html
3. www.cancer.org/cancer/managing-cancer/side-effects/fatigue-weakness-sleep.html
4. www.cancer.org/cancer/managing-cancer/side-effects/pain/peripheral-neuropathy.html
5. www.cancer.org/cancer/managing-cancer/side-effects/fertility-and-sexual-side-effects/fertility-and-women-with-cancer.html
6. www.cancer.org/cancer/managing-cancer/side-effects/fertility-and-sexual-side-effects/fertility-and-men-with-cancer.html
7. www.cancer.org/cancer/managing-cancer/treatment-types/chemotherapy.html
8. www.cancer.org/cancer/managing-cancer/side-effects.html

Last Revised: April 6, 2018

American Society of Clinical Oncology. Sarcoma, Soft Tissue: Treatment Options. 08/2017. Accessed at www.cancer.net/cancer-types/sarcoma-soft-tissue/treatment-options on April 2, 2018.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.National Cancer Institute. Adult Soft Tissue Sarcoma

Treatment (PDQ®)–Patient Version. May 15, 2017. Accessed at www.cancer.gov/types/soft-tissue-sarcoma/patient/adult-soft-tissue-treatment-pdq on April 2, 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 on April 2, 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 Nov 9;7:18.

Targeted Drug Therapy for Soft Tissue Sarcoma

- Pazopanib (Votrient)
- Tazemetostat (Tazverik)
- Other targeted drugs
- More information about targeted therapy

Targeted therapy drugs attack parts of cancer cells that make them different from normal, healthy cells. These drugs work differently from standard chemotherapy drugs, and they often have different types of side effects. 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.

As doctors learn more about the biology of sarcoma cells, targeted therapy is becoming an important treatment option for some soft tissue sarcomas.

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 people 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.

Tazemetostat (Tazverik)

Tazemetostat works by targeting EZH2, a protein known as a *methyltransferase* that normally helps some cancer cells grow. This drug can be used to treat epithelioid sarcomas that can't be removed completely by surgery. It can shrink or slow the growth of some of these cancers, although it's not yet clear if it can help people live longer.

This drug is taken as pills, typically twice a day.

Side effects

The most common side effects of this drug include pain, fatigue, nausea, vomiting, loss of appetite, and constipation. Tazemetostat can also increase the risk of developing some types of blood cancers, including certain leukemias and lymphomas.

Other targeted drugs

Many other targeted drugs might also be helpful in treating certain types of soft tissue sarcomas. Examples of these drugs include:

- **Regorafenib (Stivarga)**
- **Sorafenib (Nexavar)**
- **Sunitinib (Sutent)**
- **Larotrectinib (Vitrakvi)** (for tumors with an *NTRK* gene change)
- **Entrectinib (Rozlytrek)** (for tumors with an *NTRK* gene change)
- **Sirolimus albumin-bound nanoparticles (also known as nab-sirolimus or Fyarro)** (for advanced malignant PEComas)

Immunotherapy for Soft Tissue Sarcoma

Immunotherapy is the use of medicines to help a person's own immune system recognize and destroy cancer cells more effectively.

- Immune checkpoint inhibitors
- Autologous T cell immunotherapy
- More information about immunotherapy

Immune checkpoint inhibitors

An important part of the immune system is its ability to keep itself from attacking normal cells in the body. To do this, it uses "checkpoint" proteins on immune cells or other cells that need to be turned on (or off) to start an immune response. Cancer cells sometimes use these checkpoints to avoid being attacked by the immune system. But drugs that target these checkpoints, known as **checkpoint inhibitors**, can be used to treat some people with soft tissue sarcomas.

PD-1 and PD-L1 inhibitors

PD-1 is a checkpoint protein on immune cells called *T cells*. It normally acts as a type of "off switch" that helps keep the T cells from attacking other cells in the body. It does this when it attaches to PD-L1, a protein on some normal (and cancer) cells. When PD-1 binds to PD-L1, it basically tells the T cell to leave the other cell alone. Some cancer cells have large amounts of PD-L1, which helps them avoid being attacked by the immune cells.

Atezolizumab (Tecentriq) targets the PD-L1 checkpoint protein. By blocking PD-L1, this drug boosts the immune response against cancer cells. This can shrink some tumors or slow their growth.

Atezolizumab can be used in people with alveolar soft-part sarcoma that can't be removed with surgery or that has spread (metastasized) to other parts of the body.

This drug is given as an intravenous (IV) infusion, typically every 2, 3, or 4 weeks. It can also be given (as **Tecentriq Hybreza**) as an injection under the skin (subcutaneously) over several minutes, typically once every 3 weeks.

Pembrolizumab (Keytruda) targets the PD-1 checkpoint protein, which can also help the immune system attack the cancer cells. While this drug is not FDA-approved and sitummunoss

sarcoma cells are found to have certain gene or protein changes[1].

This drug is given as an intravenous (IV) infusion, typically every 3 or 6 weeks.

Possible side effects of checkpoint inhibitors

Side effects of these types of drugs can include fatigue, cough, nausea, itching, skin rash, loss of appetite, constipation, joint pain, and diarrhea.

Other, more serious side effects occur less often.

Infusion reactions: Some people might have an infusion reaction while getting one of these drugs. This is like an allergic reaction, and can include fever, chills, flushing of the face, rash, itchy skin, feeling dizzy, wheezing, and trouble breathing. It's important to tell your doctor or nurse right away if you have any of these symptoms while getting one of these drugs.

Autoimmune reactions: These drugs work by basically removing one of the safeguards on the body's immune system. Sometimes the immune system starts attacking other parts of the body, which can cause serious or even life-threatening problems in the lungs, intestines, liver, hormone-making glands, kidneys, or other organs.

It's very important to report any new side effects to your health care team as soon as possible. If serious side effects do occur, treatment may need to be stopped and you may get high doses of corticosteroids to suppress your immune system.

Autologous T cell immunotherapy

In this treatment, immune cells called **T cells** are removed from the patient's blood and altered in the lab to have specific receptors (called **T cell receptors**, or **TCRs**) on their surface. These receptors can attach to certain proteins on the surface of cancer cells.

The treatment is given as a one-time infusion into a vein (IV), typically about 6 weeks after the T cells are first collected. You will get chemo for several days in the week before getting this treatment, which helps it work better.

Side effects of autologous T cell immunotherapy

Because this treatment can have serious side effects, it needs to be given in a medical center that has special training with this type of treatment.

This treatment can sometimes cause **cytokine release syndrome (CRS)**, in which immune cells in the body release large amounts of chemicals into the blood. Symptoms of this life-threatening syndrome can include fever, chills, headache, nausea and vomiting, trouble breathing, very low blood pressure, a very fast heart rate, swelling, diarrhea, feeling very tired or weak, and other problems.

This treatment can sometimes cause serious **neurological (nervous system) problems**, such as confusion, trouble speaking, seizures, tremors, or changes in consciousness.

Some people might have an **infusion reaction** while getting this drug. This is like an allergic reaction, and can include fever, chills, flushing of the face, rash, itchy skin, feeling dizzy, wheezing, and trouble breathing.

Other serious side effects of this treatment can include **(Other ment c hspeci, and can include lke u**

- General body swelling

Because of the risk of side effects, you'll be watched closely at the medical center,

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 other parts of the body) 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, removing the tumor with surgery is the main treatment. Lymph nodes will also be removed if they might 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 help lower the risk of the tumor coming back.

In rare cases, amputation of part or all of a limb might be needed to remove the entire tumor, although this is now done much less often than it was in the past.

Radiation therapy with or without chemo can be used alone when the tumor's location or size or the patient's overall health makes surgery impossible.

Stage IV soft tissue sarcomas

A sarcoma is considered stage IV when it has spread to distant parts of the body. Stage IV sarcomas are rarely curable. But some patients may be cured if the main (primary) 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. In this case, the main tumors are treated as in stages II or III, and metastases are removed completely, if possible. This is still an area where doctors disagree about what the best treatment is and which patients are most likely to benefit.

For people whose primary tumor and all metastases cannot be removed completely by surgery, radiation therapy and/or chemotherapy are often used. The chemo drugs doxorubicin and ifosfamide are often the first choice — either together or along with other drugs. Gemcitabine and docetaxel may be given if the first combination doesn't work or stops working. People with angiosarcomas may benefit from treatment with

paclitaxel or docetaxel with vinorelbine.

For some types of soft tissue sarcomas, treatment with newer targeted drugs or immunotherapy might also be an option.

Recurrent sarcomas

Cancer is called **recurrent**

Last Revised: December 12, 2022

Written by

The American Cancer Society medical and editorial content team (https://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 editors and translators with extensive experience in medical writing.

cancer.org | 1.800.227.2345