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

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 Integrative Medicine

**Help getting through cancer treatment**

People with cancer need support and information, no matter what stage of illness they may be in. Knowing all of your options and finding the resources you need will help you make informed decisions about your care.

Whether you are thinking about treatment, getting treatment, or not being treated at all, you can still get supportive care to help with pain or other symptoms. Communicating with your cancer care team is important so you understand your diagnosis, what treatment is recommended, and ways to maintain or improve your quality of life.

Different types of programs and support services may be helpful, and can be 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.
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.

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.
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 Surgery¹.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects².

**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**


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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**\(^1\).

**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 **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](#)

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

**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 (Temodar®), 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 effects¹ 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 infection2 (from too few white blood cells)
• Problems with bleeding or bruising (from too few blood platelets)
• Fatigue3 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 Chemotherapy4.

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 Cancer5 and Fertility and Men With Cancer6.

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see Chemotherapy7.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects8.

Hyperlinks

1. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html
2. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/low-
blood-counts/infections.html

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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
Targeted Drug Therapy for Soft Tissue Sarcoma

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)

**More information about targeted therapy**

To learn more about how targeted drugs are used to treat cancer, see [Targeted Cancer Therapy](#).

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

**Hyperlinks**
Treatment of Soft Tissue Sarcomas, by Stage

The best chance 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, removing the tumor with surgery 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 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. 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 are most likely to benefit.

For people 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. 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). For some types soft tissue sarcomas, treatment with newer targeted drugs might also be an option.

Recurrent sarcoma

Cancer is called recurrent when it come backs 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 in a distant part of the body, chemo or targeted therapy may be used. If the sarcoma has spread only to the lungs, it may be possible to remove all the areas of spread with surgery. Radiation is often 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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