Treating Soft Tissue Sarcoma

General treatment information

Experts recommend that patients with sarcoma have a health care team made up of doctors from different specialties, such as:

- An orthopedic surgeon: a surgeon who specializes in diseases of the bones, muscles, and joints (for sarcomas of the arms and legs)
- A surgical oncologist: a doctor who treats cancer with surgery (for sarcomas of the abdomen and retroperitoneum [the back of the abdomen])
- A thoracic surgeon: a doctor who treats diseases of the lungs and chest with surgery (for sarcomas in the chest)
- A medical oncologist: a doctor who treats cancer with medicines such as chemotherapy
- A radiation oncologist: a doctor who treats cancer with radiation therapy
- A physiatrist (or rehabilitation doctor): a doctor who treat injuries or illnesses that affect how you move

Many other specialists may be involved in your care as well, including physician assistants, nurse practitioners, nurses, respiratory therapists, social workers, physical therapists, and other health professionals.

After a sarcoma is found and staged, your team will recommend one or several treatment options. This decision is important, so take time and think about all of the choices. In choosing a treatment plan, factors to consider include the type, location, and stage of the cancer, as well as your overall physical health.

The main types of treatment for soft tissue sarcoma are:

- Surgery
- Radiation
Chemotherapy
Targeted therapy

It is important to discuss all of your treatment options, including their goals and possible side effects, with your doctors to help make the decision that best fits your needs. It’s also very important to ask questions if there is anything you’re not sure about. You can find some good questions to ask in What should you ask your doctor about soft tissue sarcomas?

It also is often a good idea to seek a second opinion. A second opinion can provide more information and help you feel more confident about the chosen treatment plan. Some insurance companies require a second opinion before they will agree to pay for treatments.

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 are 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. You can also call our clinical trials matching service at 1-800-303-5691 for a list of studies that meet your medical needs, or see the Clinical Trials section on our website to learn more.

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 dangerous.

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. See the Complementary and Alternative
Help getting through cancer treatment

Your cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services are 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, support groups, 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 on call 24 hours a day, every day.

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 him or her questions about your treatment options.

Surgery for Soft Tissue Sarcomas

Depending on the site and stage of a sarcoma, surgery might be able to remove the cancer and some of the nearby tissue. 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 surrounding the tumor. 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 present at the edges, the tissue removed is said to have positive margins. This means that cancer cells may have been left behind.

When cancer cells are left after surgery, the patient may need more treatment such as radiation or another surgery. If cancer isn’t growing into the edges of the tissue removed, it is said to have negative or clear margins. The sarcoma has much less chance of coming back after surgery if it is removed with clear margins. When the tumor is in the abdomen, removing the tumor with enough normal tissue to get clear margins could be difficult because the tumor could be next to vital organs that can’t be taken out.

In the past, many of the sarcomas in the arms and legs were treated by removing the
limb (amputation). Now, this rarely is needed. Instead, most patients can be treated with surgery to remove the tumor without amputation (called limb-sparing surgery). This is usually followed by radiation therapy. These patients have the same survival rates as those who have amputations.

Sometimes, an amputation can’t be avoided. It might be the only way to remove all of the cancer. Other times, critical 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 can’t function well or would result in chronic pain, amputation may be the best option.

If the sarcoma has spread to distant sites (such as 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.

Sometimes chemotherapy (chemo), radiation, or both is given before surgery. This, called neoadjuvant treatment, can shrink the tumor and allow it to be removed completely. Chemo or radiation can also be given before surgery to treat high-grade sarcomas when there is a great risk of the cancer spreading.

Most of the time, surgery cannot cure a sarcoma once it has spread. But if it has only spread to a few spots in the lung, the metastatic tumor can sometimes be removed. This can cure many patients, or at least lead to long-term survival.

You can read more about surgery for cancer in Cancer Surgery.

- References
See all references for Soft Tissue Sarcoma

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Radiation therapy uses high-energy rays (such as x-rays) or particles to kill cancer cells.

Most of the time radiation is given after surgery as an added measure. This is called adjuvant treatment and it is done to kill any cancer cells that may be left behind after surgery.

Radiation may also be used before surgery to shrink the tumor and make the operation easier. This is called neoadjuvant treatment.

Radiation can be the main treatment for sarcoma in someone whose general health is too poor to undergo surgery.

Radiation therapy can also be used to help symptoms of sarcoma when it has spread. This is called palliative treatment.

Types of radiation therapy

External beam radiation therapy: For this treatment, radiation delivered from outside the body is focused on the cancer. 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. Often a technique called intensity modulated radiation therapy (IMRT) is used. This better focuses the radiation on the cancer and lessens the impact on healthy tissue.

In some centers, proton beam radiation is an option. 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): This is a type of external beam radiation that is available in only a few centers. 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 receives some other type of radiation after surgery.

Brachytherapy: 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 tubes) that have been placed during surgery. In high-dose rate (HDR) brachytherapy, the
pellets give off a lot of radiation in a short time, and so stay in place for only minutes at a time. In low-dose rate (LDR) brachytherapy, the pellets may stay in place for days at a time, and are then removed.

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 which area is treated and the dose given. Common side effects include

- Skin changes in the area 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 abdomen)
- Diarrhea (most common with radiation to the pelvis and abdomen)
- 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 fracture 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, headaches, and problems thinking.

If given before surgery, radiation may cause problems with wound healing.

Many side effects improve or even go away some time after radiation is finished. Some though, like bone weakness and lung damage, can be permanent.

More information on this topic can be found in [Radiation Therapy](#).

- **References**

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

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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. Chemotherapy for soft tissue sarcoma generally uses a combination of several anti-cancer drugs.

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 protects the bladder from the toxic effects of ifosfamide.

Other chemo drugs may be used as well, including cisplatin, dacarbazine (DTIC), docetaxel (Taxotere®), gemcitabine (Gemzar®), methotrexate, oxaliplatin, paclitaxel (Taxol®), vincristine, 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).

Chemotherapy drugs kill cancer cells but also damage some normal cells. Side effects depend on the type of drugs, the amount taken, and the length of treatment. Common chemo side effects 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 infection (from too few white blood cells)
- Problems with bleeding or bruising (from too few blood platelets)
• **Fatigue** and weakness (from too few red blood cells)

Most side effects disappear once treatment is stopped. Hair will grow back after treatment ends, but it might look different. There are remedies for many of the temporary side effects of chemotherapy. For example, 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 example, doxorubicin can weaken the heart if too much is given. If you are to be treated with this drug, your doctor might check your heart function with special studies before starting this drug. The doctor will also watch the dose of doxorubicin closely during therapy.

Some chemo drugs cause nerve damage (called **neuropathy**), leading to numbness, tingling, or even pain in the hands and feet. To learn more about this see [Peripheral Neuropathy Caused by Chemotherapy](#).

Chemotherapy may also permanently damage ovaries or testicles, which can lead to infertility (not being able to have children). This is discussed in more detail in [Fertility and Women With Cancer](#) and [Fertility and Men With Cancer](#).

### Isolated limb perfusion

This procedure 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 given just to that limb. Sometimes the blood is warmed up a bit to help the chemo work better (this is called **hyperthermia**). This can help shrink tumors, but it isn’t clear that it helps patients live longer than standard chemo. It should only be done at centers with a lot of experience in giving chemo this way.

• [References](#)

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

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**Targeted Therapy for Soft Tissue**
Sarcoma

Targeted therapy is a newer type of cancer treatment that uses drugs or other substances to identify and attack cancer cells while doing little damage to normal cells. These therapies attack parts of cancer cells that make them different from normal, healthy cells. Each type of targeted therapy works differently, but all alter the way a cancer cell grows, divides, repairs itself, or interacts with other cells.

Olaratumab (Lartruvo)

This drug is a type of monoclonal antibody, which is a manmade version of an immune system protein. It targets PDGFR-alpha, a protein on tumor cells that can help them grow. By blocking this protein, olaratumab can cause some tumors to shrink or stop growing. This may help people live longer.

This drug can be used along with the chemotherapy drug doxorubicin to treat soft tissue sarcomas that cannot be cured with radiation therapy or surgery.

Olaratumab is given by infusion into a vein (IV). Some people have allergic-like reactions while getting this drug, which can cause symptoms such as low blood pressure, fever, chills, and rash. Less often, reactions can be more serious or even life-threatening. Other possible side effects of this drug include nausea and vomiting, feeling tired, muscle or joint pain, swelling in the mount or throat, hair loss, headache, loss of appetite, diarrhea, and nerve damage (neuropathy), which can cause numbness, tingling, or pain in the hands or feet.

Pazopanib (Votrient)

Pazopanib blocks several cellular enzymes called tyrosine kinases that are important for cell growth and survival. In a study of patients with advanced soft tissue sarcomas that had been treated with chemotherapy, pazopanib stopped the cancers from growing for an average of about 3 months longer than the patients given a sugar pill. So far, though, this drug hasn’t been shown to help patients live longer. This drug is taken in pill form, once a day.

Common side effects include high blood pressure, nausea, diarrhea, headaches, low blood cell counts, and liver problems. In some patients this drug causes abnormal results on liver function tests, but it also rarely leads to severe liver damage that can be life threatening. Bleeding, clotting, and wound healing problems can occur, as well. This drug also rarely causes a problem with the heart rhythm or even a heart attack. If you
are taking pazopanib, your doctor will monitor your heart with EKGs as well as check your blood tests to check for liver or other problems.

**Imatinib (Gleevec)**

Imatinib is a tyrosine kinase inhibitor drug approved to treat gastrointestinal stromal tumors and some kinds of leukemia. It also can be helpful in treating desmoid tumors that can’t be removed with surgery. Although it rarely causes tumors to shrink, it often causes them to stop growing for a time, which can be very helpful.

Side effects can include mild stomach upset, diarrhea, muscle pain, and skin rashes. The stomach upset is lessened if the drug is taken with food. Imatinib can also make people retain fluid. Often this causes some swelling in the face (around the eyes) or in the ankles. Rarely the drug causes more severe problems, such as fluid build up in the lungs or abdomen or causing problems with heart function.

- References
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**Treatment of Soft Tissue Sarcomas, by Stage**

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. It is important that your surgeon and other doctors are experienced in the treatment of sarcomas. These are difficult tumors to treat and require both experience and expertise. Studies have shown that patients with sarcomas have better outcomes when they are treated at specialized cancer centers that have experience in sarcoma treatment.

**Desmoid tumors**
Desmoid tumors are often not considered soft tissue sarcomas (cancers) because although they can grow into nearby tissues and often come back after surgery, they rarely spread to distant sites.

Some desmoid tumors can be watched without treatment for a time. Treatment will be given if the tumor is growing or is causing pain or other symptoms.

If treatment is needed and the entire tumor can be removed, the first treatment is often surgery. If the entire tumor is removed and the margins are clear, no other treatment is needed. These tumors can also be treated with radiation (instead of surgery).

For tumors that are large or have come back after treatment, drug therapy may be helpful. The drug sulindac, normally used to treat arthritis, can stop tumor growth or even cause the tumor to shrink. It can take months for the drug to work, but its effect can last for years. Drugs that block estrogen (tamoxifen and toremifene) have also been helpful in some patients. Some desmoid tumors have responded to treatment with chemotherapy (chemo) using the drug doxorubicin (Adriamycin), which may be used alone or with other drugs. The combination of methotrexate and vinblastine has also been helpful. Interferon, an immune-boosting drug, has also been used with some success. Another option is the targeted drug imatinib (Gleevec).

**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 is in the head, neck, or abdomen), removing the entire tumor with enough normal tissue around it can be more difficult. 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**

Some stage III tumors have already spread to nearby lymph nodes. Most stage II and III
sarcomas are high-grade tumors. They tend to grow and spread quickly. 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 are removed (this is called local recurrence).

For all stage II and III sarcomas, surgically removing the tumor is still the main treatment. Lymph nodes will be removed as well if they contain cancer. If the tumor is large or in a place that would make surgery difficult, 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. These treatments also 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. Sometimes chemo is given as well. When chemo is given, the drug most often used is doxorubicin (Adriamycin). This drug may be combined with ifosfamide (Ifex) and other drugs.

In rare cases, amputation is needed to remove the entire tumor. As with stage I sarcomas, 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. There is evidence that chemo after surgery may benefit some people with stage II and III sarcomas.

**Stage IV soft tissue sarcoma**

A sarcoma is considered stage IV when it has spread to distant sites (M1). Stage IV sarcomas are rarely curable. But some patients may be cured if the main 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. This is still an area where doctors disagree about which patients will benefit. Those patients’ main tumors should be treated as in stages II or III, and metastases should be completely removed, if possible.

For patients whose primary tumor and all metastases cannot be completely removed by surgery, radiation therapy and/or chemotherapy are often given to relieve symptoms. The chemo drugs doxorubicin and ifosfamide are often the first choice — either alone or together with other drugs. If doxorubicin is used, it might be given along with the targeted drug olaratumab (Lartruvo). 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).

**Recurrent sarcoma**
Cancer is called *recurrent* when it comes back 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 is another option, 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 at a distant site, chemo may be given. If the sarcoma has spread only to the lungs, it may be possible to remove all the areas of spread with surgery. Radiation is used to treat sarcomas that spread to the brain, as well as any recurrences that cause symptoms such as pain.

- References

See all references for Soft Tissue Sarcoma

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