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#Reference From NCCN Guidelines

Contents

NCCN Foundation seeks to support the millions of patients and their families affected by a cancer diagnosis by funding and distributing NCCN Guidelines for Patients. NCCN Foundation is also committed to advancing cancer treatment by funding the nation's promising doctors at the center of innovation in cancer research. For more details and the full library of patient and caregiver resources, visit NCCN.org/patients.

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1
Soft tissue sarcoma basics

Soft tissue sarcoma describes a group of over 50 cancers that form in soft tissue. Soft tissue is found everywhere in the body, and includes muscle, fat, and nerves. Soft tissue sarcoma is best treated by a team of experts in different types of cancer care. This team should have experience treating your specific type of sarcoma.

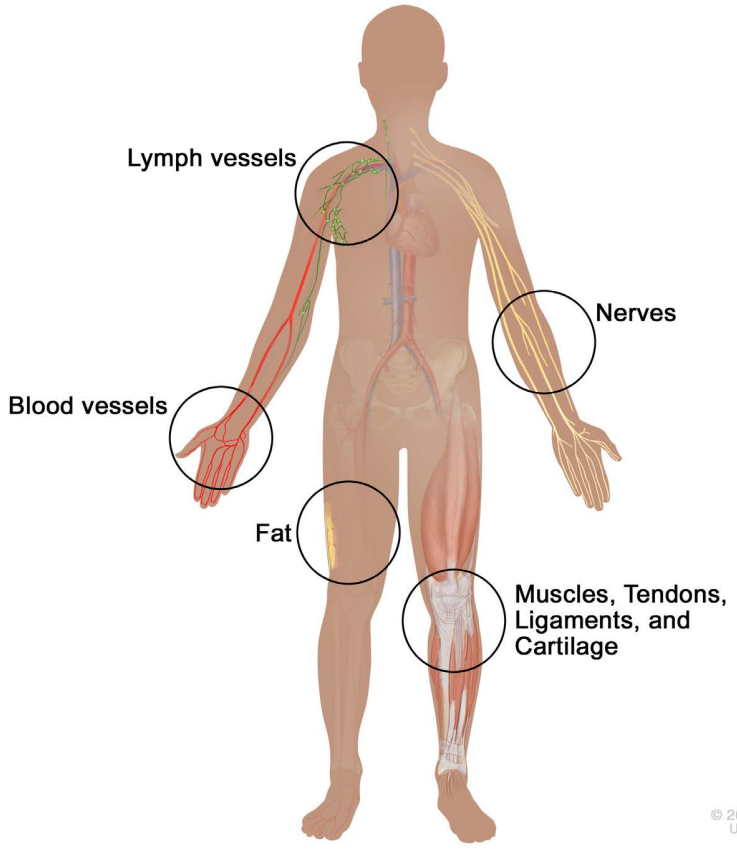
Soft tissue sarcoma starts in cells that develop into soft tissue, called mesenchymal cells. Soft tissue sarcomas form most often in the limbs (the arms and legs), followed by the torso. The torso (also called trunk) is the area of the body between the groin and the collarbone, not including the arms. The chest, abdomen, back, and pelvis are all located in the torso.

Soft tissue sarcomas can also form in the head, the neck, and the space in front of the lower spine and behind the abdominal cavity. This space is called the retroperitoneum. Organs in the retroperitoneum include the adrenal glands, aorta, kidneys, esophagus, ureters, pancreas, rectum, and parts of the stomach and colon.

The other, less common type of sarcoma is bone sarcoma, such as osteosarcoma. Soft tissue sarcoma is the focus of this book.

Soft tissue sarcoma

Soft tissue includes fat, muscles, nerves, tendons, blood and lymph vessels, and other supportive tissues of the body.



Types of soft tissue sarcoma

The cells of each type of sarcoma look different under a microscope. Physicians with specialty training in pathology (pathologists) examine tissue using a microscope to determine the type of soft tissue sarcoma. An accurate tumor type (also called histology or histopathology) is needed to create the best treatment plan for you.

There are over 50 types of soft tissue tumors, which behave very differently. Some may return to the same area but not spread further, while others often spread to other parts of the body. This is one reason why it is important to have an accurate tumor type.

Most soft tissue tumors fall into one of the following categories:

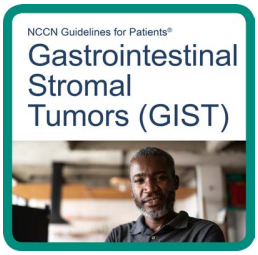
- Adipocytic tumors
- Fibroblastic/myofibroblastic tumors
- Fibrohistiocytic tumors
- Peripheral nerve sheath tumors
- Smooth muscle tumors
- Skeletal muscle tumors
- Tumors of uncertain differentiation
- Vascular tumors

Undifferentiated pleomorphic sarcoma (UPS) is the most common type of soft tissue sarcoma. In UPS, the cells don't look like the soft tissue in which they are found. This is one example of a tumor in the "uncertain differentiation" category. These are most common in the legs and arms.

Liposarcomas are fat-forming tumors. These are most common in the retroperitoneum and legs.

Leiomyosarcomas are tumors made of smooth muscle. These are most often found in the abdomen.

Gastrointestinal stromal tumors (GIST) form in the digestive tract. GIST is addressed separately in the *NCCN Guidelines for Patients: Gastrointestinal stromal tumors (GIST)* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Soft tissue sarcomas in this book are divided into:

- Sarcomas in the limbs, outer torso, head, or neck
- Sarcomas inside the torso
- Desmoid tumors (aggressive fibromatosis)
- Rhabdomyosarcoma (RMS)
- Atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDLS)

The importance of specialized care

Soft tissue sarcomas are rare. Some are so rare that there aren't specific recommendations for how to treat them. Most doctors are not experts in diagnosing or treating these tumors. It is important to find a treatment center or hospital that has experts with experience in your type of soft tissue sarcoma. Ideally you would receive all your care with those experts. For those who live far from such centers, it often is possible for the experts to provide you with a treatment plan and coordinate with your local doctors. This would allow you to receive care closer to home. Certain types of treatments, such as clinical trials or specialized radiation therapy, may only be available at specialized treatment centers or hospitals.

Risk factors

A small number of people carry a mutation (change) in their genes that increases the risk of developing soft tissue sarcoma. Those with such mutations are said to have hereditary or genetic syndromes. For soft tissue sarcomas, these include:

- Hereditary retinoblastoma
- Neurofibromatosis
- Li-Fraumeni syndrome
- Hereditary non-polyposis colorectal cancer (HNPCC)/Lynch syndrome
- Familial adenomatous polyposis (FAP) and Gardner syndrome
- Carney-Stratakis syndrome (increases the risk of GIST)

We inherit our genes from our parents. In hereditary or genetic syndromes, your siblings, your parents' siblings, and your grandparents

Adolescents and young adults (AYAs) with cancer have unique medical, mental, emotional, and social concerns.

The focus of this guide is treatment of soft tissue sarcoma in adults. AYAs with soft tissue sarcoma have age-specific problems and needs. See the NCCN Guidelines for Patients: Adolescent and Young Adult Cancer for more information.

often have the same mutation. It is important that you share what you know about your family history with your doctor. This helps them determine if you may benefit from genetic testing. If so, a genetic counselor may speak to you about the results. A genetic counselor is an expert who has special training in genetic diseases. More information on genetic testing is provided in the next chapter.

Key points

- Soft tissue sarcoma starts in cells that develop into soft tissue, called mesenchymal cells.
- Soft tissue includes fat, muscles, nerves, tendons, and blood and lymph vessels.
- There are over 50 types of soft tissue sarcoma that can be found anywhere in the body.
- Diagnosis and treatment are based on the location and type of soft tissue sarcoma.
- It is important to find a treatment center or hospital that has experts with experience in your type of soft tissue sarcoma.
- Certain genetic syndromes may put you at risk for developing soft tissue sarcoma. You may be referred for genetic counseling.

2
Testing

Testing for suspected soft tissue sarcoma starts with imaging scans and a biopsy. Other testing is individualized based on the location of the tumor and your health history.

Imaging

Imaging tests make pictures of areas inside of the body. They can show the primary (first) tumor and other possible areas of cancer. The imaging tests used most often for soft tissue sarcoma are computed tomography (CT) and magnetic resonance imaging (MRI).

CT takes many x-rays of the same body part from different angles. Using computer technology, the images are combined to make detailed pictures.

MRI uses radio waves and powerful magnets to take pictures of areas inside the body. It does not use radiation.

The choice of CT versus MRI depends on the type and location of the tumor. For example, MRI is preferred for tumors in the brain, spine, and extremities. Both CT and MRI can provide details about the size of the tumor, and how close it is to nearby organs, nerves, and blood vessels.

A radiologist is an expert at interpreting medical scans. A radiologist will review the images and write a report. The radiologist will send this report to your doctor who will discuss the results with you. Depending on what they see, the radiologist may suggest that you have another or different type of imaging scan.

PET

Positron emission tomography (PET) scans use a radioactive substance called a tracer or isotope. This substance identifies tissues that are using a lot of energy. Most cancers, including sarcomas, use much more energy

CT scan

A CT scan is a more detailed type of x-ray. It is painless and noninvasive. CT takes many images from different angles. A computer combines the images to make 3D pictures.

than normal cells and show up as brights spots on PET scans. Not all bright spots are cancer. PET is usually combined with CT scans and can provide additional helpful information.

Contrast

Before CT or MRI, you may be given a contrast agent. Contrast is a liquid put into your body that changes how certain types of tissue appear when imaged. Often, your doctors will order the same type of scan (CT or MRI) with and without contrast to get better information about the tumor. Tell your care team if you have had a reaction to contrast in the past. If you have, you might be given medicine, such as Benadryl or prednisone. Contrast might not be used if you have a serious allergy or if your kidneys don't work well.

Biopsy

After imaging tests, your doctor will usually order or perform a biopsy. This involves removing a sample of the tumor for testing. The preferred type of biopsy for soft tissue sarcoma is a **core needle biopsy.** It removes tissue with a wide, hollow needle.

Another type called an incisional or open biopsy removes a small amount of tissue through a cut in the skin or body. Sometimes, doctors may perform a biopsy using a thinner needle, called fine needle aspiration (FNA).

Most soft tissue sarcomas are deep within the body. For these tumors, biopsies are guided using imaging, such as an ultrasound or CT. Image-guided needle biopsies are usually needed for tumors in the chest, abdomen, pelvis, and retroperitoneum. If your doctor believes your tumor is close to the skin, they may do a biopsy without imaging guidance.

Testing the tumor sample

A pathologist is an expert in examining tissue and cells to diagnose disease. After examining the tissues and cells with a microscope, the pathologist writes a summary of the results. This is called a pathology report.

The pathologist often performs other tests to see if the tumor cells have specific genes or proteins. This is often necessary to diagnose the tumor type. This may also help choose the best treatment plan for your tumor type. Because not all pathologists are familiar with every type of soft tissue sarcoma, review at an experienced center is recommended. Ask questions about your biopsy results and what it means for your treatment.

Sometimes the pathologist is not able to make a diagnosis from your biopsy. This may be because the tissue is dead, because the biopsy does not have enough cells for all of the tests needed, or because no cancer is seen.

Genetic tests

Certain genetic syndromes increase the risk of developing a soft tissue sarcoma. Mutations (changes) in different genes cause each of these genetic syndromes. Testing for gene mutations that cause these syndromes may be helpful for treating soft tissue sarcoma, and for monitoring your or your family members for cancers.

Share what you know about your family history with your care team. If your health care provider thinks you may have a genetic syndrome that is causing your cancer, they might refer you for genetic testing. If needed, testing should be carried out by a pathologist who is experienced in genetic testing techniques. A genetic counselor may speak to you about the results. A genetic counselor is an expert who has special training in genetic diseases.

Hereditary retinoblastoma

Retinoblastoma is cancer that forms in the retina (the light-sensitive layers of nerve tissue at the back of the eye). It is most common in children under the age of 5. It may be hereditary or nonhereditary. The hereditary form increases the risk of leiomyosarcoma and osteosarcoma.

Li-Fraumeni syndrome

Families with Li-Fraumeni syndrome have a mutation in the *TP53* gene. This gene helps control repair or survival of damaged cells. Families with Li-Fraumeni syndrome have a history of sarcoma, breast cancer, adrenocortical tumors, and some brain tumors. Those who have a personal and/or family history that suggests Li-Fraumeni syndrome should be considered for further genetic testing.

FAP

People with familial adenomatous polyposis (FAP) have an adenomatous polyposis coli (APC) gene mutation. This syndrome causes many polyps to form in the colon and rectum. The polyps start as noncancerous growths, but over time they can turn into cancer.

FAP has a related syndrome called Gardner syndrome. In addition to bowel polyps, those with Gardner syndrome have abnormal bony growths and skin and soft tissue tumors.

Your care team will gather information to determine if you are likely to have Gardner syndrome or FAP. Both are associated with desmoid tumors. If you are diagnosed with a desmoid tumor, your family history should be looked at closely. Your doctor may suggest a colonoscopy.

Neurofibromatosis

This rare genetic condition causes tumors to grow on nerves. These tumors are usually malignant peripheral nerve sheath tumors (MPNSTs). About 5 out of 100 people with neurofibromatosis will develop soft tissue sarcoma.

Lynch syndrome

Lynch syndrome is also called hereditary non-polyposis colorectal cancer (HNPCC). Those born with Lynch syndrome are at high risk of developing colorectal cancer and some other cancers, particularly endometrial and ovarian cancer. While not common, Lynch syndrome can also increase the risk of soft tissue sarcoma.

Carney-Stratakis syndrome

This rare inherited disorder causes gastrointestinal stromal tumors (GIST) and paragangliomas. Paragangliomas are tumors that form in embryonic nervous tissues in the head, neck, and torso. They are usually benign.

Key points

- Testing for suspected soft tissue sarcoma starts with imaging and a biopsy.
- A core needle biopsy is generally preferred for diagnosing soft tissue sarcoma.
- MRI and CT scans provide details about the size and location of the tumor.
- Other testing is individualized based on the location of the tumor and your health history.
- Your health care provider might refer you for genetic testing to learn more about your cancer.

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NCCN.org/patients/comments

3 Treatments

Your treatment options will depend on the location and type of soft tissue sarcoma. This chapter describes surgery, radiation therapy, and other commonly used treatments.

Treatment can be local, can work throughout the body ("systemic"), or can be a combination of both. Local therapy treats the tumor directly. Surgery and radiation therapy are local therapies. Chemotherapy, targeted therapy, and immunotherapy are systemic therapies.

Surgery

Surgery, also called resection, is an operation to remove cancer from the body. It is the primary (main) treatment for sarcoma. Imaging tests are used to determine if your cancer is resectable (can be removed with surgery) or unresectable (cannot be safely removed with surgery). Unresectable tumors often involve other critical body structures. Or they are so extensive that surgery cannot remove the whole tumor.

The method and extent of surgery for sarcoma depend on the tumor location and size, and whether it involves nearby body parts. The goal of surgery is to remove all the cancer. To do so, the tumor is removed along with a rim of normal-looking tissue around its edge. The edges of the tissue that is resected are called the surgical margins.

Your surgeon will use the pre-surgery imaging studies to plan the operation and will explain what to expect for recovery afterward. The surgeon will attempt to remove all of the sarcoma, but sometimes tumor cells that could not be seen during the operation are left behind.

A clear or negative margin (R0) is the best result. It means that there was a layer of normal surrounding the entire tumor that was removed. It also means there is a decreased chance that any cancer cells were left behind.

If all of the visible tumor is removed but the pathologist finds microscopic cancer cells at the margin, it is called an R1 positive margin. If the surgeon is unable to remove all of the visible tumor, it is called an R2 positive margin.

Sometimes it is not possible to safely remove the sarcoma with a cancer-free margin. If this is expected, your surgeon may place clips in your body after removing the tumor. The clips will mark where to give radiation therapy. You might have more than one surgery. You might also have a wound drain to prevent fluid from collecting in the body after surgery.

Limb-sparing

When surgery involves a limb (an arm or a leg), the goal is to have a functional or working limb after the tumor is removed. This is called limb-sparing surgery. Most surgeries to remove sarcomas in a limb use limb-sparing surgery.

Rehabilitation will likely be part of a limb-sparing treatment plan. Occupational therapy helps with daily life skills. Physical therapy helps your body move and function.

After surgery to remove a sarcoma, some people may suffer from lymphedema, or swelling in the limb. This can worsen if radiation is also a part of treatment. Lymphedema therapists can provide

techniques and stockings that can help reduce this swelling.

Amputation

Amputation is the removal of a limb or other body part. A surgeon who is an expert in soft tissue sarcoma should be consulted to determine if amputation is necessary or if limb salvage is possible.

Rehabilitation, such as physical and occupational therapy, will be part of this treatment. Seek a peer support group to connect with others who have experienced the loss of a limb. This will aid in your recovery and help you to maintain an active lifestyle.

Metastasectomy

A metastasectomy is surgery to remove one or more distant (metastatic) tumors. It may be an option when the primary tumor can be completely removed and there are only a few small metastatic tumors that can be safely removed by surgery. Location, the amount of tissue that must be removed, and your overall health are factors in determining if you are eligible for metastasectomy.

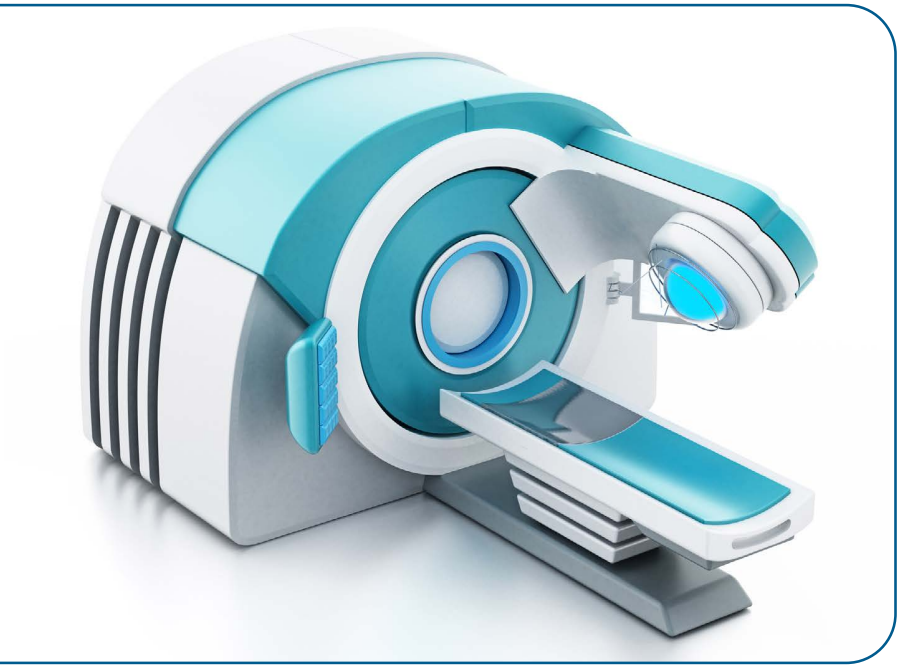
A metastasectomy may be done at the same time as surgery to remove the primary tumor. Or it may be done during a separate operation. The amount of time needed for the surgery and recovery depends on many factors. Some factors include the size, location of the metastases, and the type of surgery needed.

Palliative surgery

Surgery may also be used to relieve symptoms caused by the cancer or to extend life. This is called palliative surgery.

Radiation therapy

Radiation therapy uses high-energy radiation from x-rays, gamma rays, protons, and other sources to kill cancer cells and shrink tumors. It is also used to treat pain.



Radiation therapy

Radiation therapy uses high-energy radiation from x-rays, gamma rays, protons, and other sources to kill cancer cells. It is given alone or with chemotherapy over a fixed period of time.

Radiation may be used to shrink the tumor before surgery. This can make it easier to resect (remove) the tumor, but may increase wound healing problems.

Radiation is sometimes given to kill leftover cancer cells after surgery. This may be recommended if not all cancer was removed during surgery, or if there is a high risk of recurrence. When given after surgery, radiation can lower the risk of the tumor growing back. It may lead to increased scarring or stiffness (fibrosis), which can reduce movement in the affected limb.

Radiation can also be used during surgery, called intraoperative radiation therapy (IORT), or to ease pain or discomfort caused by the tumor. This is called palliative radiation therapy.

The 2 main types of radiation treatment are explained next. The method used depends on the size and type of sarcoma, and the goal of treatment.

EBRT

External beam radiation therapy (EBRT) uses a machine outside of the body to aim radiation at the tumor. Types of EBRT used to treat soft tissue sarcoma include:

- **Intensity-modulated radiation therapy (IMRT)** is an advanced radiation technique that uses many small beams of different strengths.
- **Image-guided radiation therapy (IGRT)** helps guide the radiation beam during treatment. Tumors can shift slightly within the body and can change shape and size between and during treatment visits. IGRT can improve how well IMRT targets the tumor.
- **Stereotactic body radiation therapy (SBRT)** uses high-energy radiation beams to treat metastatic tumors in five or fewer sessions.
- **Proton beam radiation therapy (PBRT)** uses streams of particles called protons to kill tumor cells and may reduce the amount of radiation damage to nearby healthy tissue.
- **Intraoperative radiation therapy (IORT)** uses radiation treatment aimed directly at the tumor during surgery.

Brachytherapy

In brachytherapy, a radiation source is placed inside or next to the tumor. This is also called internal radiation therapy. Brachytherapy may be used alone or with EBRT. When used with EBRT it is called a brachytherapy boost.

LDR brachytherapy

Low dose-rate (LDR) brachytherapy uses thin, hollow needles to place radioactive seeds into the tumor. The seeds are about the size of a grain of rice. They are guided into the tumor with imaging tests.

The seeds usually contain radioactive iodine or palladium. They will stay in the tumor and give a low dose of radiation for a few months. The radiation will travel a very short distance. This will allow for a large amount of radiation within a small area while sparing nearby healthy

Treatment team

Treating soft tissue sarcoma takes a team approach. Treatment planning should involve a team of doctors from different fields of medicine who have experience in your type of soft tissue sarcoma. Some members of your care team will be with you throughout treatment, while others will only be there for parts of it. Get to know your care team and let them get to know you.

- **A pathologist** diagnoses a tumor by studying cells and tissues under a microscope.
- **A diagnostic radiologist** reads the results of x-rays and other imaging tests such as an MRI or CT scan.
- **An orthopedic surgical oncologist** performs operations to remove soft tissue and bone sarcomas that involve the pelvis, spine, and extremities. They perform limb salvage surgery and reconstruct bones after a bone tumor is removed.
- **A surgical oncologist** performs operations to remove cancer from the torso, abdomen, and sometimes the extremities.
- **A medical oncologist** treats cancer in adults using systemic therapy. This person often leads the overall treatment team.
- **A radiation oncologist** prescribes and plans radiation therapy to treat cancer.

Depending on your diagnosis, your team might include:

- **Oncology nurses** provide hands-on care, like giving systemic therapy, answering questions, and helping with side effects.
- **Advanced practice providers** are nurse practitioners and physician assistants who work with physicians to provide care.
- **An interventional radiologist** performs needle biopsies of tumors and sometimes ablative therapies.
- **A thoracic surgeon** removes tumors from within the chest or lungs.
- **A plastic surgeon** works with other surgeons to help with reconstruction of soft tissues after tumor removal.
- **Physical and occupational therapists** help people recover from surgery and build strength, mobility, and resume daily tasks.
- **A dietitian or nutritionist** helps meet your nutrition needs during treatment

Keep a list of names and contact information for each member of your team. This will make it easier for you and anyone involved in your care to know who to contact with questions or concerns.

tissue. Over time, the seeds will stop radiating, but will stay in your body (permanent).

HDR brachytherapy

High dose-rate (HDR) brachytherapy uses thin needles placed inside the tumor. These needles are then attached to tubes called catheters. Radiation is delivered through these catheters. After treatment, the needles and catheters are removed.

Other local treatments

Isolated limb infusion

Isolated limb infusion/perfusion is used to treat sarcoma of the limb. It is a type of regional limb therapy. It involves delivering anti-cancer drugs directly into an arm or leg. During surgery, the flow of blood is stopped for a short period of time while the drugs are injected into the blood of the limb (infusion). This limits the effect of chemotherapy on the rest of the body. This option should only be done at centers or hospitals with experience in regional limb therapy.

Ablation

Ablation uses extreme cold or extreme heat to destroy cancer cells. It can destroy small tumors with little harm to nearby tissue.

There are 2 types of thermal ablation used to destroy cancer cells:

- Cryotherapy kills cancer cells by freezing them with a very cold substance.
- Radiofrequency ablation (RFA) kills cancer cells by heating them with high-energy radio waves.

Both types use a special needle, called a probe, which is inserted into the tumor.

With cryotherapy, a medical gas is passed through the probe to cause below-freezing temperatures. This freezes the tumor to destroy it. With radiofrequency ablation, the probe emits radio waves to heat the tumor and destroy it.

The probe can be guided into place with a CT scan, ultrasound, or other imaging tests. The probe is removed when treatment is done.

Embolization

Embolization treats tumors by cutting off their blood supply. It is most commonly used before surgery to remove a tumor. A catheter is inserted into an artery and guided to the tumor. Once in place, beads are inserted to block the blood flow. In chemoembolization, the beads are coated with chemotherapy. Radioembolization uses small radioactive beads.

Systemic therapy

A cancer treatment that affects the whole body is called systemic. Chemotherapy, targeted therapy, and immunotherapy are types of systemic therapy. Many systemic therapies are liquids that are injected into a vein or under the skin with a needle. Others are taken by mouth as a pill.

Chemotherapy

Chemotherapy kills fast-growing cells throughout the body, including cancer cells and normal cells. There are many chemotherapy drugs that might be used to treat soft tissue sarcoma. More than one drug may be used.

Most chemotherapy is given in cycles of treatment days followed by days of rest. This allows the body to recover between cycles. Cycles vary in length depending on which

drugs are used. The number of treatment days per cycle and the total number of cycles given also vary.

Chemoradiation

Treatment that combines chemotherapy with radiation therapy is called chemoradiation. Chemotherapy may improve how well radiation works, and that is why they are sometimes used together. It is a combination of systemic and local therapies.

Targeted therapy

Targeted therapy is drug therapy that focuses on specific or unique features of cancer cells. Targeted therapy isn't used for every sarcoma. It might be used alone or with chemotherapy.

Immunotherapy

The immune system is the body's natural defense against infection and disease. It is a complex network of cells, tissues, and organs. Immunotherapy increases the activity of your immune system. By doing so, it improves your body's ability to find and destroy cancer cells. Pembrolizumab, ipilimumab, and nivolumab are types of immunotherapy.

Clinical trials

A clinical trial is a type of medical research study. After being developed and tested in a laboratory, potential new ways of fighting cancer need to be studied in people. If found to be safe and effective in a clinical trial, a drug, device, or treatment approach may be approved by the U.S. Food and Drug Administration (FDA).

Everyone with cancer should carefully consider all of the treatment options available for their cancer type, including standard treatments and clinical trials. Talk to your doctor about whether a clinical trial may make sense for you.

Phases

Most cancer clinical trials focus on treatment. Treatment trials are done in phases.

- **Phase 1** trials study the dose, safety, and side effects of an investigational drug or treatment approach. They also look for early signs that the drug or approach is helpful.
- **Phase 2** trials study how well the drug or approach works against a specific type of cancer.
- **Phase 3** trials test the drug or approach against a standard treatment. If the results are good, it may be approved by the FDA.
- **Phase 4** trials study the long-term safety and benefit of an FDA-approved treatment.

Who can enroll?

Every clinical trial has rules for joining, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, or general health. These requirements ensure that participants are alike in specific ways and that the trial is as safe as possible for the participants.

Informed consent

Clinical trials are managed by a group of experts called a research team. The research team will review the study with you in detail, including its purpose and the risks and benefits of joining. All of this information is also provided in an informed consent form.

Read the form carefully and ask questions before signing it. Take time to discuss it with family, friends, or others you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

Start the conversation

Don't wait for your doctor to bring up clinical trials. Start the conversation and learn about all of your treatment options. If you find a study that you may be eligible for, ask your treatment team if you meet the requirements. If you have already started standard treatment, you may not be eligible for certain clinical trials. Try not to be discouraged if you cannot join. New clinical trials are always becoming available.

Frequently asked questions

There are many myths and misconceptions surrounding clinical trials. The possible benefits and risks are not well understood by many with cancer.

Will I get a placebo?

Placebos (inactive versions of real medicines) are almost never used alone in cancer clinical trials. It is common to receive either a placebo with a standard treatment or a new drug with a standard treatment. You will be informed, verbally and in writing, if a placebo is part of a clinical trial before you enroll.

Are clinical trials free?

There is no fee to enroll in a clinical trial. The study sponsor pays for research-related costs, including the study drug. You may, however, have costs indirectly related to the trial, such as the cost of transportation or child care due to extra appointments. During the trial, you will continue to receive standard cancer care. This care is billed to—and often covered by—insurance. You are responsible for copays and any costs for this care that are not covered by your insurance.

Finding a clinical trial

In the United States

NCCN Cancer Centers
NCCN.org/cancercenters

The National Cancer Institute (NCI)
cancer.gov/about-cancer/treatment/clinical-trials/search

Worldwide

The U.S. National Library of Medicine (NLM)
clinicaltrials.gov

Need help finding a clinical trial?

NCI's Cancer Information Service (CIS)
1.800.4.CANCER (1.800.422.6237)
cancer.gov/contact

Key points

Surgery

- When possible, surgery is often the preferred treatment for soft tissue sarcoma.
- Surgery removes the tumor along with some normal-looking tissue around it (the surgical margin).

Radiation therapy

- Radiation therapy is a local treatment that is often used before or after surgery to lower the risk of the tumor growing back after surgery.
- It can also be used alone as a main (primary) treatment, or to relieve pain and discomfort caused by the tumor(s).
- The two main types of radiation therapy are external beam radiation therapy (EBRT) and brachytherapy.
- Intensity-modulated radiation therapy (IMRT), image-guided radiation therapy (IGRT), proton therapy, intraoperative radiation therapy (IORT), and stereotactic body radiation therapy (SBRT) are types of EBRT.
- SBRT uses high-energy radiation beams to treat metastatic tumors in five or fewer sessions.
- If radiation therapy is planned, ask your doctor if proton therapy is an option.

Other local treatments

- Other local treatments include isolated limb infusion/perfusion, ablation, and embolization.

Systemic therapy

- Systemic therapy is the use of substances to treat cancer cells throughout the body.
- Chemotherapy, targeted therapy, and immunotherapy are types of systemic therapy.

Clinical trials

- A clinical trial is a type of research that studies an investigative treatment to see how safe it is and how well it works.

4

Sarcomas in the limbs, outer torso, head, or neck

Sarcomas in the limbs, outer torso, head, or neck

This chapter describes treatment options for soft tissue sarcomas in the arms, legs, outer torso, head, and neck. Treatment is based on the type and location of the tumor, as well as the cancer stage.

Many types of soft tissue sarcoma can form in the head, neck, limbs, or outer torso. The limbs include the arms, legs, and thighs. The outer torso includes the chest, back, buttocks, and abdomen.

Head and neck soft tissue sarcomas are similar to sarcomas in other parts of the body. But they can be hard to treat because of the location. The tumor is likely to be close to bones, muscles, and nerves, making surgery more difficult. Areas of the head and neck that could be affected include:

- Neck and throat such as the larynx and pharynx
- Face and scalp
- Sinus and nasal (nose) cavities
- Mouth and lips

Some types of soft tissue sarcoma found in the limbs, outer torso, or head and neck are listed alphabetically below.

- **Alveolar soft part sarcoma (ASPS)** begins in the soft tissue that surrounds organs and other tissues. It commonly occurs in the thigh, but can be found in the arms, legs, or torso.
- **Angiosarcoma** begins in the cells that line blood vessels or lymph vessels. These often occur in the limbs but can occur anywhere.
- **Dermatofibrosarcoma protuberans** occurs in the layer of tissue right under the skin. It starts as a nodule or thickening that grows slowly.
- **Epithelioid sarcoma** begins as a slow-growing, firm lump in the deep soft tissue or skin of the arms, hands, or fingers. It may also occur in the legs, torso, or head and neck. It is usually painless but tends to spread and return after treatment.
- **Leiomyosarcoma** is cancer of smooth muscle cells that can arise almost anywhere in the body.
- **Myxoid/round cell liposarcoma** begins in fat cells. It usually occurs in the thigh but can be found in the outer torso and buttocks.
- **Atypical lipomatous tumors (ALT)/ well-differentiated liposarcoma (WDLS)** have cells that look like fat cells under a microscope. They tend to grow more slowly than poorly differentiated or undifferentiated cancer cells. When these grow in the extremities, they are not considered cancerous.

Stage 1A/1B

A stage 1 sarcoma in the outer torso or limbs may be small or large in size. Cancer has not spread to nearby lymph nodes or to distant sites. Stage 1 is further grouped into stage 1A and stage 1B. These stages are low-grade. This means they are not expected to grow quickly.

Surgery is the recommended treatment for stage 1A/1B sarcomas in these areas. If there is a serious risk of complications from surgery, shrinking the tumor with radiation therapy first may be recommended.

The goal of surgery is to remove the sarcoma with a cancer-free surgical margin (R0). However, to avoid cutting nerves and blood vessels, your surgeon may not be able to remove a large enough surgical margin. In this case, there is a higher chance that cancer cells will be left behind. Using a microscope, a pathologist will examine the surgical margin for cancer cells.

If the sarcoma was completely resected (R0), you can start follow-up care.

If not enough tissue around the sarcoma was removed (R1 or R2), you may have a second surgery or radiation therapy. Radiation therapy is a very good option for stage 1B tumors. For stage 1A tumors, observation is also an option. Observation is a period of scheduled follow-up testing to watch for signs of cancer spread or return.

Surgery has risks

Those with some types of soft tissue sarcoma have a greater risk of complications from surgery. Also, sarcomas in the face, head, or neck might be more difficult to remove. Ask your doctor about the risks of surgery, what will be removed, and what this means in terms of your function and recovery.

Follow-up care

A physical exam is recommended every 3 to 6 months for the first 2 to 3 years after treatment. After that, visits are spaced out to once per year.

Your doctor will consider an imaging study after treatment to serve as your baseline. You may have periodic imaging of the surgery site. This is on a case-by-case basis, based on the risk of recurrence. If the tumor is in an area that is easy to see and examine, imaging may not be needed.

If cancer is likely to spread to your lungs, you may get scans of your chest.

After surgery to remove a soft tissue sarcoma, you should be evaluated for rehabilitation services. Such services include occupational and physical therapy. Occupational therapy can help with daily life skills. Physical therapy helps your body move and function.

Stage 2, 3, and some stage 4

The following information applies to stage 2 and stage 3 sarcomas in the limbs or outer torso. It also applies to stage 4 sarcomas that have spread to nearby lymph nodes, but not to distant sites. There is no distant metastasis.

Treatment for these stages is based on whether surgery can be done. If the tumor can be safely removed and you will have good use of the limb or area, it is considered resectable.

Surgery might not be the first treatment. The tumor may be too large, or removing it may limit the use of a limb or other body part. Surgery may also cause health-related problems. Therefore, treatment is grouped into:

- Resectable with good function,
- Resectable with poor function, and
- Unresectable.

All options include surgery to remove the tumor. Other treatment is often given before or after surgery.

Treatment before surgery may include chemoradiation, radiation therapy, or systemic therapy. The goal is to shrink the tumor before surgery.

Resectable with good function

Stage 2

Surgery is the main treatment for resectable stage 2 sarcomas. Radiation therapy may be given before or after surgery.

When given first, radiation can reduce the tumor size and improve the outcome of surgery. When given after surgery, radiation can lower the risk of the cancer returning. It may also improve how well your limb works after surgery. One downside is that, if done before surgery or before the surgical wound has fully healed, it will likely slow healing of the wound.

Stage 3 and some stage 4

Possible treatment pathways for stage 3 and non-metastatic stage 4 sarcomas include:

- Surgery → radiation therapy or chemoradiation
- Radiation therapy → surgery → possibly systemic therapy
- Chemoradiation → surgery → possibly systemic therapy
- Systemic therapy → surgery → radiation therapy or chemoradiation

If systemic therapy is planned, the drug(s) used will depend on the tumor type. Preferred systemic therapies for non-specific types of soft tissue sarcoma are listed in **Guide 1**.

Preferred systemic therapies for specific tumor types are listed in **Guide 2**.

Alveolar soft part sarcoma (ASPS), atypical lipomatous tumor/well-differentiated liposarcoma (ALT/WDLS), and clear cell sarcomas do not usually respond to chemotherapy.

Unresectable or resectable with poor function

Surgery may not be a first treatment option for stage 2, 3, or non-metastatic stage 4 sarcomas. This is often due to the tumor location and/or size. One of the below treatments is usually used first to shrink the tumor. If it works well, surgery may be possible.

- Radiation therapy
- Chemoradiation
- Systemic therapy
- Isolated limb infusion

After treatment, your doctors will assess if you are able to have surgery with good results. Good results include a surgical margin larger than 1 cm, cancer-free margins, and good use of your limb or other body part. This means a tumor that was unresectable is now resectable.

If the cancer becomes resectable, see treatment information under “Resectable with good function” on page 26.

If the cancer becomes resectable but surgery would cause a loss of function, either radiation therapy or radical surgery (eg, amputation) may be recommended.

If the cancer **remains unresectable** after treatment, options include:

- Radiation therapy (if area was not already treated)
- Systemic therapy
- Palliative surgery
- Observation (if no symptoms)
- Best supportive care

Amputation is the removal of a limb or other body part. Amputation may be considered if a limb will not work after surgery. A surgeon who is an expert in soft tissue sarcoma should be consulted to determine if amputation is necessary or if limb salvage is possible.

If the cancer is causing symptoms, palliative surgery or other best supportive care may be recommended for relief from symptoms.

Follow-up care

A physical exam is recommended every 3 to 6 months for the first 2 to 3 years after treatment. For the following 2 years, exams are recommended every 6 months. After that, visits are spaced out to once per year. After 10 years, the risk of recurrence is very low. Talk to your treatment team about how long you should have testing.

Your doctor will order imaging when treatment ends. These scans will serve as a baseline to compare against future imaging results. You will continue to get scans of the site where the primary tumor was. How often depends on how likely the cancer is to return.

Imaging of the chest is also recommended to look for cancer spread for these stages.

After surgery to remove a soft tissue sarcoma, you should also be evaluated for rehabilitation services. Such services include occupational and physical therapy. Occupational therapy can help with daily life skills. Physical therapy helps your body move and function.

Guide 1
Preferred systemic therapy for non-specific types of soft tissue sarcoma

For before or after surgery	• AIM (doxorubicin, ifosfamide, mesna) • Ifosfamide, epirubicin, mesna
As first systemic therapy for advanced or metastatic disease	Anthracycline-based regimens: • Doxorubicin • Epirubicin • Liposomal doxorubicin • AD (doxorubicin, dacarbazine) • AIM • Ifosfamide, epirubicin, mesna For *NTRK* gene fusion-positive sarcomas only: • Larotrectinib • Entrectinib
As next-in-line therapy for advanced or metastatic disease	• Pazopanib • Eribulin (especially for liposarcoma) • Trabectedin (especially for liposarcoma and leiomyosarcoma)

At this time, the systemic therapies listed in this guide are preferred for general soft tissue sarcoma. There are other recommended drug therapies that may be a better fit for you. Talk to your care team about your options.

Guide 2 Preferred systemic therapy regimens for specific types of soft tissue sarcoma	
Alveolar soft part sarcoma (ASPS)	Sunitinib Pazopanib Pembrolizumab, with or without axitinib Atezolizumab
Angiosarcoma	Paclitaxel Anthracycline- or gemcitabine-based regimens recommended for general soft tissue sarcoma types
DFSP with fibrosarcomatous transformation	Imatinib
Epithelioid sarcoma	Tazemetostat
Extraskeletal osteosarcoma	Usually treated with Ifosfamide or platinum-based therapy (cisplatin/doxorubicin)
Dedifferentiated chordoma	Anthracycline- or gemcitabine-based regimens recommended for general soft tissue sarcoma types
Inflammatory myofibroblastic tumor (IMT) with *ALK* translocation	Alectinib Brigatinib Ceritinib Crizotinib Lorlatinib
Malignant PEComa	Albumin-bound sirolimus
Solitary fibrous tumor	Bevacizumab and temozolomide Sunitinib Sorafenib Pazopanib
Tenosynovial giant cell tumor/ pigmented villonodular synovitis	Pexidartinib
Recurrent angiomyolipoma, lymphangioleiomyomatosis	Sirolimus Everolimus Temsirolimus
At this time, the systemic therapies listed in this guide are preferred for specific types of soft tissue sarcoma. There may be other recommended options that may be a better fit for you. Talk to your care team about your options.	

Metastatic stage 4

In stage 4 soft tissue sarcoma in the limbs or outer torso, the cancer may be in lymph nodes, distant areas, or both. Sarcomas do not often spread to lymph nodes.

The following information applies to sarcomas found in distant areas when first diagnosed. Treatment depends on the extent of the cancer.

If there are only a few small metastatic tumors that can be treated with local therapies, treatment according to “Resectable with good function” on page 26 is recommended. Your treatment team will consider the following options for treating the metastatic tumors:

- Systemic therapy (see **Guides 1** and **2**)
- Metastasectomy, with or without radiation therapy
- Stereotactic body radiation therapy (SBRT)
- Ablation methods
- Embolization (not for lung tumors)
- Observation

Follow-up care

A physical exam is recommended every 2 to 6 months for the first 2 to 3 years after treatment. For the following 2 years, exams are recommended every 6 months. After that, visits are spaced out to once per year. After 10 years, the risk of recurrence is very low. Talk to your treatment team about how long you should have testing.

Imaging of the chest and of any areas where cancer has spread is recommended.

Your doctor may order imaging when treatment ends. These scans will serve as a baseline to compare against future imaging results. You may continue to get scans of the site where the primary tumor was. How often depends on how likely the cancer is to return.

Widespread cancer

If the metastases are widespread, the goal of care is palliative. Palliative care does not try to cure the cancer. It is used to relieve pain and discomfort, to reduce symptoms caused by the cancer, and to extend life. Palliative treatment options include:

- Systemic therapy (see **Guides 1** and **2**)
- Radiation (SBRT)
- Surgery
- Observation, if asymptomatic
- Supportive care
- Ablation procedures
- Embolization (not for lung tumors)

Systemic therapy, radiation therapy, and surgery may reduce cancer symptoms by stopping or slowing tumor growth. Other options include ablation and embolization.

Supportive care is treatment to relieve symptoms of cancer and side effects from treatment. Supportive care is not cancer treatment. It might include pain relief, emotional or spiritual support, financial aid, or family counseling. Tell your care team how you are feeling and about any side effects.

Follow-up care, including imaging, for widespread metastatic disease is individualized. Talk with your doctor about how often you may need imaging and what other care to expect.

Recurrence

A recurrence is the return of cancer after a cancer-free period. Treatment options depend on the location of the new growth. If sarcoma returns to the area where it started, it is called local recurrence. Local recurrence is often treated as if it were newly diagnosed.

If the cancer returns and spreads to areas far from where it started, it is called distant recurrence. The cancer is metastatic. Treatment for metastatic disease is based on whether the new cancer growth is:

- Only in one organ
- Only in a specific region or found in lymph nodes
- Widespread

If the cancer has **spread to only one organ** and the tumor is small, surgery to remove the metastatic tumor (metastasectomy) is often recommended, especially for lung tumors. Systemic therapy and possibly also radiation therapy may be given before or after surgery.

Other recommended options for recurrence in one organ include:

- Radiation (SBRT) with or without systemic therapy
- Ablation procedures
- Embolization procedures (not for lung tumors)
- Observation

If cancer returns to the same general area and is **found in lymph nodes**, a regional lymph node dissection is recommended. This is surgery to remove lymph nodes from your body. It might be followed by radiation therapy.

If radiation therapy is planned, systemic therapy may also be given.

The cancer may return to areas far from where it started. If the cancer is **widespread**, the main goal of care is not to cure the cancer. The aim is to control its growth, extend your life, and make you more comfortable. This care could include:

- Systemic therapy (see **Guides 1** and **2**)
- Destroying metastatic tumors with radiation therapy (SBRT)
- Surgery
- Observation (if you don't have symptoms)
- Supportive care
- Ablation procedures
- Embolization procedures (not for lung tumors)

Key points

Stage 1A/1B

- Most stage 1 sarcomas in the limbs, outer torso, head, or neck can be treated with surgery.
- If there is a serious risk of complications from surgery, shrinking the tumor with radiation therapy first may be recommended.
- If tumor cells remain after surgery, you may have a second surgery or radiation therapy. Observation is also an option for stage 1A tumors. Radiation therapy is a very good option for stage 1B tumors.

Stage 2, 3, and some stage 4

- Surgery is the main treatment for resectable stage 2 sarcomas. Radiation therapy may be given before or after surgery.
- For stage 3 and non-metastatic stage 4 sarcomas, surgery may come first, followed by radiation therapy or chemoradiation. Or radiation therapy and/or systemic therapy may be given first to shrink the tumor.
- If surgery is not possible, other treatment is given first. This could include radiation therapy, chemoradiation, systemic therapy, or isolated limb infusion.
- If the limb will not function after surgery, amputation is sometimes the best option. Your treatment team will consider advances in limb reconstruction.

Metastatic stage 4

- If there are only a few small metastatic tumors, treatment according to "Resectable with good function" on page 26 is recommended.
- Local therapies and/or systemic therapy will be considered to treat the metastatic tumors. Observation may also be an option.
- The goal of care for widespread metastatic cancer is to control its growth and improve comfort and quality of life.
- Follow-up care, including imaging, for widespread metastatic disease is individualized.

Recurrence

- Recurrence is the return of cancer after a disease-free a period.
- Treatment for recurrence is based on the extent and location of the cancer.
- Local recurrence is often treated as if it were newly diagnosed.
- Metastasectomy is often recommended if cancer is found in one organ and the metastatic tumor is small.
- A lymph node dissection is recommended if cancer is found in any lymph nodes.

5
Sarcomas inside the torso

Soft tissue sarcomas found in organs inside the chest, abdomen, or pelvis are usually treated with surgery. Together these areas are called the inner trunk or torso.

The **retroperitoneum** is the area in the back of the abdomen. Organs found in the retroperitoneum include the adrenal glands, aorta, kidneys (and ureters), part of the pancreas, rectum, and parts of the stomach and colon.

Liposarcoma and leiomyosarcoma are the most common types of soft tissue sarcoma found in the retroperitoneum.

Tumors in the retroperitoneum tend to grow very large before causing symptoms. They tend to push against or invade nearby structures such as veins and arteries. The size of the tumor and how close it is to major blood vessels may make surgery difficult.

A biopsy is needed to confirm soft tissue sarcoma in the retroperitoneum.

The organs of the **abdomen and chest**, such as the lungs and the bowel, are known as visceral organs. They include any soft organ, but not lymph nodes.

Resectable

When possible, surgery is recommended for soft tissue sarcomas inside the torso. For sarcomas thought likely to return and/or spread, treatment may be given before surgery. This could involve radiation therapy or systemic (chemotherapy, immunotherapy, or targeted) therapy.

If treatment before surgery is being considered, an image-guided core needle biopsy is needed. Otherwise, a biopsy may not be performed. An experienced sarcoma pathologist should review the biopsy samples.

If systemic therapy before surgery is planned, the drug(s) used will depend on the subtype of soft tissue sarcoma. Preferred systemic therapies for non-specific types of soft tissue sarcoma are listed in **Guide 1** on page 28. Preferred systemic therapies for specific tumor types are listed in **Guide 2** on page 29.

After surgery, a pathologist examines the surgical margin for cancer cells. Depending on the results, more treatment may be needed. For any sarcoma thought to be at high risk of spreading, systemic therapy will be considered after surgery.

The best possible surgical outcome is called complete resection or negative margin resection. Doctors call this **R0**. It means that no cancer cells were found in the surgical margin. If achieved, radiation therapy is **not** needed after surgery. For tumors close to vital organs, veins, arteries, and other tissues, complete resection is not always possible.

If the surgeon removes all of the visible tumor but microscopic cancer cells remain in the margin, it is an **R1** positive margin. An **R2** positive margin means that all of the visible tumor could not be removed. This can

happen when the tumor is close to important structures, like large blood vessels or nerves. A small number of those with R1 or R2 surgery results may benefit from radiation therapy after surgery.

If the tumor was not totally removed (R2 positive margin), a second surgery to remove the remaining cancer may be an option. This is generally only considered if complete resection is thought to be possible. You should also be healthy enough to have the surgery.

Follow-up care

When treatment is over, testing is used to look for signs of recurrence. Testing includes physical exams and imaging. For imaging, computed tomography (CT) is generally preferred for sarcomas in these areas. MRI is also a recommended option. In some cases, positron emission tomography (PET) may be combined with CT.

Imaging and a physical exam are recommended every 3 to 6 months for the first 2 to 3 years after treatment. For the following two years, testing is spaced to every 6 months. After that, testing is recommended once per year.

If the cancer is likely to spread to the lungs, you may get imaging tests of your chest.

Unresectable or stage 4

A tumor that cannot be safely removed with surgery is called unresectable. These include tumors that involve vital structures and tumors whose removal could cause complications or death. Overall health also plays a role. You should also be healthy enough to have major abdominal surgery.

Stage 4 tumors can be any size (T) and any grade (G). Cancer has spread to nearby lymph nodes (this is not common) or to distant areas (M1).

A biopsy will be done first to stage the tumor. An experienced sarcoma pathologist should review any biopsy and tissue samples.

If the tumor is slow-growing and not causing symptoms, observation without treatment is one recommended option. Sometimes an unresectable tumor can become resectable. An attempt might be made to shrink the tumor using systemic therapy and/or radiation therapy. Imaging tests will be used to see if the tumor has shrunk and if surgery is now possible.

If the tumor becomes resectable, see “Resectable” on page 34. If the tumor stays unresectable or gets worse, options include:

- Trying a different systemic therapy
- Palliative radiation therapy
- Palliative or best supportive care

Palliative care does not try to cure the cancer; it aims to relieve your discomfort. Systemic therapy or radiation therapy may reduce cancer symptoms by stopping or slowing tumor growth.

Recurrence

The return of cancer after a cancer-free period is called recurrence.

If the cancer is resectable, surgery is recommended. After surgery, you may have radiation therapy or systemic therapy.

In some cases, radiation therapy or systemic therapy is given before surgery. Systemic therapy before surgery will only be considered if there is a high risk of spread. It is not recommended for low-grade tumors.

If the new cancer growth cannot be removed with surgery, see “Unresectable or stage 4” on the previous page.

Key points

Resectable

- When possible, surgery is recommended for soft tissue sarcomas inside the torso.
- For tumors expected to return and/or spread, radiation therapy or systemic (drug) therapy may be given first.
- If surgery removes all of the cancer, it is called complete resection (R0) or negative margin resection. Radiation therapy is not needed.
- If microscopic cancer cells remain or the tumor could not be fully removed, radiation may be considered after surgery.

Unresectable or stage 4

- Observation without treatment is a recommended option if the tumor is slow-growing and not causing symptoms.
- Systemic therapy and/or radiation therapy may be used to try to shrink the tumor.
- If the tumor becomes resectable, surgery is recommended.
- If the tumor stays unresectable or grows, options include trying a different systemic therapy, palliative radiation therapy, and best supportive care.

Recurrence

- If the new cancer growth is resectable, surgery is recommended.
- Radiation therapy or systemic therapy may be given after surgery.
- In some cases, radiation therapy or systemic therapy is given before surgery.

6
Desmoid tumors

Desmoid tumors are persistent, non-cancerous growths. They are treated like cancer, even though they do not spread through the body. These tumors are usually observed or treated with systemic therapy.

Also known as aggressive fibromatosis or desmoid-type fibromatosis, desmoid tumors aren't considered cancerous because they do not spread in the body. But they can be persistent in the area where they start. If they recur (return), they return to the same area as the primary (first) tumor.

Testing

Your doctor will ask about your health history and perform a physical exam. A sample of the tumor will also be removed for testing. This is called a biopsy. Imaging of the tumor may also be needed. Either computed tomography (CT) or magnetic resonance imaging (MRI) is recommended.

Hereditary syndromes

Desmoid tumors often occur for unknown reasons. However, some rare syndromes increase the risk of cancer and desmoid tumors. Familial adenomatous polyposis (FAP) can cause hundreds to thousands of polyps to form in the colon and rectum. The polyps start as noncancerous growths, but over time they can turn into cancer. In addition to bowel polyps, those with Gardner syndrome have abnormal bony growths and skin and soft tissue tumors. Gardner syndrome is a type of FAP. Your care team will gather information to determine if you are likely to have Gardner syndrome or FAP.

Observation

Many newly-diagnosed desmoid tumors can be observed without treatment. If the tumor is stable and not causing pain, observation is often an option. The tumor is monitored with imaging (CT or MRI). Imaging is recommended every 3 months to start. Your imaging schedule will depend on the location of the tumor, the risk of progression, and any symptoms of progression. More frequent imaging may be needed if you have symptoms or if the tumor is near important body structures.

If the tumor is causing pain or other symptoms, drug therapy may be an option. This depends on where the tumor is in the body and the safety of the drug.

If the desmoid tumor worsens somewhat but your symptoms aren't bad, your doctor may recommend continuing observation. However, if growth continues and causes severe symptoms or problems, treatment is needed. See "Progression" on the next page.

Large tumors that are causing problems will be treated based on the location of the tumor, if it is causing pain, or if it interferes with movement.

Progression

Treatment for desmoid tumors often includes systemic therapy and/or ablation. Surgery and radiation therapy are less commonly used. Each of these is discussed in more detail next. The best treatment(s) for you will depend on the location of the tumor.

Systemic therapy

There are different types of systemic therapy used to treat desmoid tumors. Options include chemotherapy, tyrosine kinase inhibitors (TKIs), and a gamma secretase inhibitor (GSI). Recommended systemic therapies for desmoid tumors are listed in **Guide 3**.

Treatment might include nonsteroidal anti-inflammatory drugs (NSAIDs). NSAIDs help reduce fever, swelling, pain, and redness. Sulindac (Clinoril) and celecoxib (Celebrex) are NSAIDs that are commonly prescribed for people with symptoms. Celecoxib can cause heart issues or put those with heart issues at greater risk for side effects.

Ablation

Ablation uses extreme cold or extreme heat to destroy desmoid tumors. It can help control the size of the tumor. Types of ablation include radiofrequency ablation (RFA), cryotherapy, and high-intensity focused ultrasound (HIFU). These procedures are typically done at specialty hospitals or health care centers that specialize in treating desmoid tumors.

Guide 3
Systemic therapy for desmoid tumors

Drug name	How it's given	Type of drug
Nirogacestat (Ogsiveo)	pill	targeted therapy
Sorafenib (Nexavar)	pill	targeted therapy
Methotrexate and vinorelbine	infusion	chemotherapy
Methotrexate and vinblastine	infusion	chemotherapy
Imatinib (Gleevec)	pill	targeted therapy
Liposomal doxorubicin (Doxil)	infusion	chemotherapy
Doxorubicin with or without dacarbazine	infusion	chemotherapy
Pazopanib (Votrient)	pill	targeted therapy

Surgery

Desmoid tumors are hard to completely remove. They also frequently grow back after surgery. For these reasons, surgery is generally not a first-line treatment, unless agreed upon by a team of experts.

Surgery might not be considered if there is risk of serious complications. It may be too dangerous based on the tumor location or your overall health.

If surgery is planned and the results are very good, it is called complete resection. Doctors write this as R0. Observation is recommended.

If microscopic cancer cells remain after surgery, it is called R1. Options may include observation, another surgery, and radiation therapy to kill leftover cancer cells.

If much of the cancer could not be removed, it is called R2. Options may include:

- Radiation therapy
- Systemic therapy
- Ablation procedures
- Observation

If these treatments fail, radical surgery may be considered. Radical surgery could involve removing large areas of tissue and/or organs affected by the tumor. However, a main goal of treatment is to maintain function. Amputation is almost never considered.

Radiation therapy

In general, radiation therapy is only recommended for desmoid tumors in the limbs (arms and legs), outer torso, head, neck, abdominal wall, pelvis, trunk, and chest cavity. It is not recommended for desmoid tumors in the abdomen or retroperitoneum, except under special circumstances.

The retroperitoneum is the area in the back of the abdomen. Organs in the retroperitoneum include the adrenal glands, aorta, kidneys (and ureters), part of the pancreas, rectum, and parts of the stomach and colon.

Follow-up care

After treatment, you will be monitored for symptoms. A change in symptoms may be a sign of tumor growth. Report any new or worsening symptoms to your doctor. Follow-up tests are used to see if the desmoid tumor is growing or has returned.

Imaging

Imaging with CT or MRI every 3 to 6 months for 2 to 3 years is recommended. After that, imaging is spaced out to once or twice per year. Depending on the location of the tumor, ultrasound may also be used for long-term follow-up.

Rehabilitation

You should receive rehabilitation services if needed. This may include occupational and/or physical therapy.

Key points

- Desmoid tumors (also called aggressive fibromatoses) are persistent, non-cancerous growths.
- These tumors do not spread through the body but are treated when they cause symptoms in the affected area.

Observation

- Many newly-diagnosed desmoid tumors can be observed without treatment. The tumor is monitored with imaging tests.
- If the tumor is causing pain or other symptoms, drug therapy may be an option.
- If the cancer grows and causes severe symptoms or problems, treatment for progression is needed.

Progression

- Treatment for desmoid tumors could include systemic therapy, surgery, radiation therapy, and/or ablation.
- Surgery usually isn't the first treatment, unless agreed upon by a team of experts.
- In general, radiation therapy is not recommended for desmoid tumors in the abdomen or retroperitoneum.
- Follow-up care will include rehabilitation, if needed. This may include occupational or physical therapy.
- You will have tests to see how the tumor is responding to treatment. Tests include a medical history, physical exam, and imaging.

Let us know what you think!

Please take a moment to complete an online survey about the NCCN Guidelines for Patients.

NCCN.org/patients/response

7
Rhabdomyosarcoma (RMS)

Rhabdomyosarcoma (RMS) is more common in children than adults. This type of soft tissue sarcoma usually starts in cells that develop into skeletal muscle.

Rhabdomyosarcoma (RMS) is a rare type of sarcoma of the skeletal muscle. Skeletal muscles are those that we can control or move. RMS can also be found in parts of the body that don't normally have skeletal muscle.

Before we are born, cells called rhabdomyoblasts begin to form. These cells eventually become skeletal muscle. But sometimes rhabdomyoblasts develop into RMS. RMS is more common in children because it starts in rhabdomyoblasts.

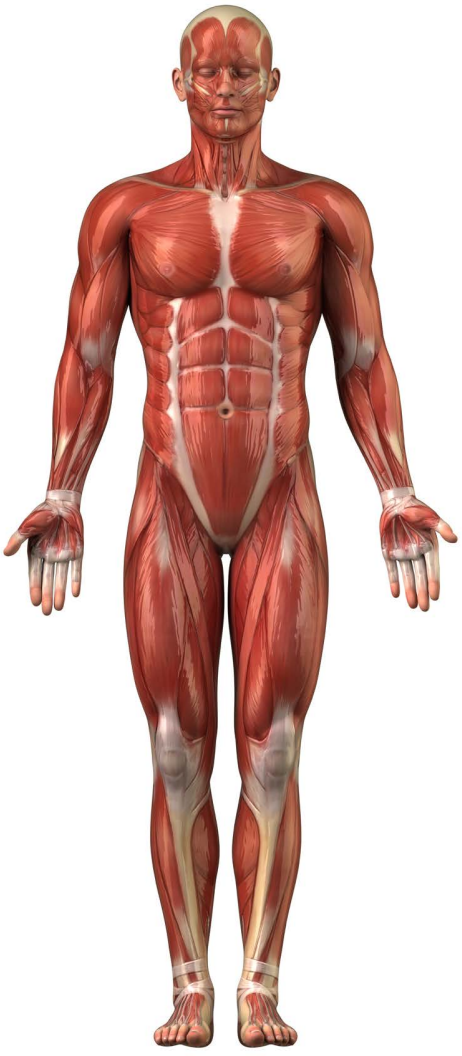
Risk factors for RMS include Li-Fraumeni syndrome and neurofibromatosis type 1 (NF1).

RMS is divided into pleomorphic and non-pleomorphic. Non-pleomorphic RMS is more common and usually affects children. It includes the embryonal, alveolar, and spindle cell/sclerosing subtypes.

Pleomorphic RMS usually affects adults and is rare.

Skeletal muscles

Skeletal muscles are usually attached to bones of our skeleton. These are muscles we can control or move.



Non-pleomorphic RMS

This type of RMS is mostly seen in children. It includes the following subtypes:

- Alveolar
- Embryonal
- Spindle cell/sclerosing

Alveolar RMS (or ARMS) is often found in skeletal muscle of the limbs, but can be found in the torso, head, and neck. It is more aggressive than embryonal RMS (or ERMS). ERMS tends to occur in the head, neck, bladder, vagina, or in or around the prostate and testicles.

Spindle cell/sclerosing includes tumors with *VGLL2*-related fusions, intraosseous (bone) tumors, and tumors with *MYOD1* mutation. The aggressiveness of the tumor is directly related to the mutation present.

Spindle cell/sclerosing rhabdomyosarcomas that are present at birth or that occur shortly after birth often are less aggressive than other rhabdomyosarcomas.

Seek treatment at a cancer center with expertise in the management of pediatric cancers, including non-pleomorphic RMS.

Treatment

Seeking treatment at a cancer center experienced in treating non-pleomorphic RMS is strongly recommended. Treatment planning should involve a pediatric oncologist as well as medical, radiation, and surgical oncologists.

Treatment often involves a combination of surgery, systemic therapy, and radiation therapy.

At this time, the following chemotherapy regimens are preferred for systemic therapy:

- VAC regimen: vincristine, dactinomycin, and cyclophosphamide
- VAI regimen: vincristine, dactinomycin, ifosfamide

If chemotherapy with one of these regimens works very well, sometimes maintenance chemotherapy is given to maintain the good results. This depends on the aggressiveness of the tumor.

If treatment with the VAC or VAI regimen is not possible, there are other recommended options for systemic therapy.

Pleomorphic RMS

This rare type of RMS occurs mainly in adults and tends to grow quickly. The most common areas in which pleomorphic RMS forms are the limbs, torso wall, reproductive system, or urinary system. The urinary system includes the kidneys, bladder, urethra, and ureter.

The prognosis (outlook) for pleomorphic RMS is typically poor. This type is usually excluded from RMS and soft tissue sarcoma clinical trials. This means there is little data on the best treatment for adults with RMS.

At this time, pleomorphic RMS is treated like high-grade undifferentiated pleomorphic sarcoma (UPS). Treatment may involve surgery, radiation therapy, and systemic therapy.

Key points

Overview

- Rhabdomyosarcoma (RMS) is a rare type of sarcoma of the skeletal muscle.
- RMS can also be found in parts of the body that don't normally have skeletal muscle.
- RMS is more common in children because it starts in premature cells called rhabdomyoblasts.
- Risk factors for RMS include Li-Fraumeni syndrome and neurofibromatosis type 1.

Non-pleomorphic RMS

- RMS in children is called non-pleomorphic RMS. It includes embryonal, alveolar, and spindle cell/sclerosing subtypes.
- Treatment should be planned by a team of specialists at a center experienced in treating non-pleomorphic RMS.
- Those with RMS often require more than one type of cancer treatment including surgery, radiation therapy, and/or systemic therapy.
- Either the VAC or the VAI regimen is preferred for chemotherapy. Maintenance chemotherapy may be given.

Pleomorphic RMS

- This type of RMS occurs mainly in adults and tends to grow quickly.
- Pleomorphic RMS is treated like high-grade undifferentiated pleomorphic sarcoma (UPS).
- Treatment may involve surgery, radiation therapy, and systemic therapy.

8

Atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDLS)

Liposarcoma is a rare type of soft tissue sarcoma that starts in fat cells. Atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDLS) is one of five major types of liposarcoma.

This complex name reflects tumors that look the same through the microscope but act differently depending on where they form.

Tumors that develop in the arms or legs may return to the same area after a very long time (years to decades). They rarely develop dedifferentiation (explained below) and even more rarely cause death. Some sarcoma doctors prefer the name “ALT” for these tumors, since they are not very aggressive.

Tumors that develop in the retroperitoneum often return to the same area, often develop dedifferentiation (see below), and eventually (years to decades later) cause death. Most sarcoma doctors prefer the name “WDLS” for these more aggressive tumors.

While rare, over time ALT/WDLS can transform into a related type of liposarcoma called dedifferentiated liposarcoma. The features signaling this change can be diagnosed by a pathologist who examines tissue with a microscope, or in imaging studies.

Unlike ALT/WDLS, dedifferentiated liposarcoma may be fast-growing. It is also more likely to return after treatment and/or spread to distant areas of the body (metastasize).

Treatment

Limbs, abdominal wall, or torso

Treatment for ALT/WDLS that forms in these areas depends on whether there is evidence of dedifferentiation. If there isn’t evidence of dedifferentiation or higher-grade sarcoma, surgery alone to remove the tumor is recommended.

After surgery, tumor cells may remain in the normal tissue surrounding the tumor. But more treatment is not typically needed because these tumors do not usually spread within the body. If the tumor returns, either observation or another surgery is recommended.

Radiation therapy is not recommended for atypical lipomatous tumors because they are considered benign. See “Follow-up” on the next page for information on surveillance after surgery.

If there are signs of dedifferentiation, the same treatment used for other soft tissue sarcomas that form in these areas is recommended. See Part 4 of this guide, “Sarcomas in the limbs, outer torso, head, or neck.”

Retroperitoneum or scrotum

For WDLS that forms in the retroperitoneum or scrotum, treatment is not guided by whether there is dedifferentiation. The same treatment used for other soft tissue sarcomas that form in these areas is recommended. See Part 5 of this guide, “Sarcomas inside the torso.”

Follow-up

Surveillance after treatment includes physical exams and imaging of the area where the tumor formed. Suggested schedules for these tests are provided below. But you may need them more or less often. This will depend on the risk of the cancer returning.

A physical exam is recommended every 6 to 12 months for the first two years after treatment. After that, one exam per year is recommended.

How often imaging is needed will be individualized. Your doctor will consider the location (how easily it can be examined without imaging) and how likely the cancer is to return. Imaging of the chest to look for cancer spread is not recommended unless dedifferentiation was present.

We want your feedback!

Our goal is to provide helpful and easy-to-understand information on cancer.

Take our survey to let us know what we got right and what we could do better.

NCCN.org/patients/feedback

9
Making treatment decisions

It's important to be comfortable with the cancer treatment you choose. This choice starts with having an open and honest conversation with your care team.

It's your choice

In shared decision-making, you and your doctors share information, discuss the options, and agree on a treatment plan. It starts with an open and honest conversation between you and your doctor.

Treatment decisions are very personal. What is important to you may not be important to someone else.

Some things that may play a role in your decision-making:

- What you want and how that might differ from what others want
- Your religious and spiritual beliefs
- Your feelings about certain treatments
- Your feelings about pain or side effects
- Cost of treatment, travel to treatment centers, and time away from school or work
- Quality of life and length of life
- How active you are and the activities that are important to you

Think about what you want from treatment. Discuss openly the risks and benefits of specific treatments and procedures. Weigh options and share concerns with your doctor.

If you take the time to build a relationship with your doctor, it will help you feel supported when considering options and making treatment decisions.

Second opinion

It is normal to want to start treatment as soon as possible. While cancer can't be ignored, there is time to have another doctor review your test results and suggest a treatment plan. This is called getting a second opinion, and it's a normal part of cancer care. Even doctors get second opinions!

Things you can do to prepare:

- Check with your insurance company about its rules on second opinions. There may be out-of-pocket costs to see doctors who are not part of your insurance plan.
- Make plans to have copies of all your records and imaging studies sent to the doctor you will see for your second opinion.

Support groups

Many people diagnosed with cancer find support groups to be helpful. Support groups often include people at different stages of treatment. Some people may be newly diagnosed, while others may be finished with treatment. If your hospital or community doesn't have support groups for people with cancer, check out the websites listed in this book.

Questions to ask

Possible questions to ask your doctors are listed on the following pages. Feel free to use these questions or come up with your own. Be clear about your goals for treatment and find out what to expect from treatment.

Questions about cancer testing

1. What tests will I have?
2. Do the tests have any risks?
3. Will my insurance pay for all of the tests you are recommending?
4. Do I need to do anything to prepare for testing?
5. Should I bring someone with me to the appointments?
6. Where do I go for testing, and how long will it take?
7. If any of the tests will hurt, what will you do to make me comfortable?
8. How soon will I know the results and who will explain them to me?
9. How can I get a copy of the pathology report and other test results?
10. Is there an online portal with my test results?

Questions about treatment options

1. What are my treatment options?
2. Is a clinical trial an option for me?
3. What will happen if I do nothing?
4. Are you suggesting options other than what NCCN recommends? If yes, why?
5. How do my age, sex, overall health, and other factors affect my options?
6. What if I am pregnant, or planning to become pregnant?
7. Does any option offer a cure or long-term cancer control?
8. What are the side effects of the treatments?
9. How do I get a second opinion?
10. How long do I have to decide about treatment, and is there a social worker or someone who can help me decide?

Questions about what to expect

1. Does this hospital or cancer center offer the best treatment for me?
2. Do I have a choice of when to begin treatment? How long will treatment last?
3. Will my insurance cover the treatment you're recommending?
4. Are there any programs to help pay for treatment?
5. What supportive care and services are available to me and my caregivers?
6. Who should I contact with questions or concerns if the office is closed?
7. How will you know if treatment is working?
8. What are the chances of the cancer worsening or returning?
9. What follow-up care is needed after treatment?
10. Will I be able to work while I am undergoing treatment?
11. If I need to take time off of work, how much time would this be?
12. Will I need any accommodations to continue to work during treatment?

Questions about side effects

1. What are the possible complications and side effects of treatment?
2. Does the cancer itself cause any side effects?
3. Which side effects are most common and how long do they usually last?
4. Which side effects are serious or life-threatening?
5. Are there any long-term or permanent side effects?
6. What symptoms should I report right away, and who do I contact?
7. What can I do to prevent or relieve the side effects of treatment?
8. Do any medications worsen side effects?
9. Do any side effects lessen or worsen in severity over time?
10. Will you stop or change treatment if there are serious side effects?

Questions about clinical trials

1. Do you recommend that I consider a clinical trial for treatment?
2. How do I find clinical trials that I can participate in?
3. What are the treatments used in the clinical trial?
4. Has the treatment been used for other types of cancer?
5. What are the risks and benefits of this treatment?
6. What side effects should I expect and how will they be managed?
7. How long will I be in the clinical trial?
8. Will I be able to get other treatment if this doesn't work?
9. How will you know if the treatment is working?
10. Will the clinical trial cost me anything?

Questions about resources and support

1. Who can I talk to about help with housing, food, and other basic needs?
2. What help is available for transportation, childcare, and home care?
3. How much will I have to pay for treatment?
4. What help is available to pay for medicines and treatment?
5. What other services are available to me and my caregivers?
6. How can I connect with others and build a support system?
7. How can I find in-person or online support?
8. Who can help me with my concerns about missing work or school?
9. Who can I talk to if I don't feel safe at home, at work, or in my neighborhood?
10. How can I get help to stop smoking or vaping?

Resources

Clear Cell Sarcoma Foundation
Clearcellsarcoma.org

Desmoid Tumor Research Foundation
DTRF.org

Leiomyosarcoma Support & Direct Research Foundation
lmsdr.org

National Cancer Institute (NCI)
cancer.gov/types

National LeioMyoSarcoma Foundation
nlmsf.org

Northwest Sarcoma Foundation
nwsarcoma.org

Osteosarcoma Institute
osinst.org

Sarcoma Coalition
sarcomacoalition.us

Sarcoma Foundation of America
curesarcoma.org

The Life Raft Group
liferaftgroup.org

The Paula Takacs Foundation
paulatakacsfoundation.org

Triage Cancer
Triagecancer.org

U.S. National Library of Medicine Clinical Trials Database
clinicaltrials.gov/

Take our survey, and help make the NCCN Guidelines for Patients better for everyone!

NCCN.org/patients/comments

Words to know

ablation
A treatment that destroys very small tumors with heat, cold, lasers, or chemicals. Also called ablative therapy.

biopsy
A procedure that removes fluid or tissue samples to be tested for disease.

brachytherapy
A treatment with radiation from an object placed near or in the tumor. Also called internal radiation.

chemoembolization
A cancer treatment with chemotherapy-coated beads that block blood supply to tumors.

chemoradiation
Treatment with a combination of chemotherapy and radiation therapy.

chemotherapy
Drugs that kill cancer cells by damaging or disrupting the making of the genetic code.

computed tomography (CT)
A test that uses x-rays from many angles to make a picture of the insides of the body.

contrast
A substance put into your body to make clearer pictures during imaging tests.

core needle biopsy
A procedure that removes tissue samples with a hollow needle. Also called core biopsy.

cryotherapy
A type of ablation therapy that kills cancer cells by freezing them.

Desmoid tumor
A mass of fibrous cells that grows into nearby tissue and rarely spreads to distant sites (often not cancer). Also known as aggressive fibromatosis.

distant recurrence
Cancer that has come back after treatment and is found in a part of the body far from the first (primary) tumor.

embolization
A treatment that cuts off blood supply to tumors with beads inserted into an artery.

external beam radiation therapy (EBRT)
A cancer treatment with radiation delivered from a machine outside the body.

familial adenomatous polyposis (FAP)
A hereditary condition that causes many abnormal growths (polyps) to form in the bowel. Increases the risk of sarcoma.

Gardner syndrome
A hereditary syndrome that causes bowel polyps, abnormal bony growths, and skin and soft tissue tumors. A type of familial adenomatous polyposis (FAP).

grade
A rating of how quickly a cancer is expected to grow. Based on how abnormal the cancer cells look under a microscope.

histology
The structure of cells, tissue, and organs as viewed under a microscope.

immunotherapy
A treatment with drugs that help the body find and destroy cancer cells.

intensity-modulated radiation therapy (IMRT)
Treatment with radiation that uses small beams of different strengths based on the thickness of the tissue. A type of EBRT.

intraoperative radiation therapy (IORT)
Radiation therapy given during surgery.

isolated limb infusion/perfusion
A method of giving cancer drugs directly into the bloodstream of a limb (arm or leg).

Li-Fraumeni syndrome
A health condition passed down in a family that increases the chance of getting sarcoma and other cancers.

local recurrence
Cancer that returns to the same general area after treatment.

lymph node
A small, bean-shaped, disease-fighting structure.

lymph node dissection
Surgery to remove one or more lymph nodes.

magnetic resonance imaging (MRI)
A test that uses radio waves and powerful magnets to make pictures of the insides of the body.

medical oncologist
A doctor who's an expert in cancer drugs.

metastasectomy
Surgery to remove tumors that formed far from the first site of cancer. Used to reduce cancer burden and to ease symptoms.

metastasis
The spread of cancer cells from the first (primary) tumor to a new site.

neurofibromatosis
A rare genetic condition that causes tumors to grow on nerves. Increases the risk of soft tissue sarcoma.

observation
A period of testing for changes in cancer status while not receiving treatment.

occupational therapist
An expert in helping people live life unaided.

oncology surgeon
A doctor who's an expert in operations that remove cancer.

palliative care
Health care that includes symptom relief but not cancer treatment. Also sometimes called supportive care.

pathologist
A doctor who's an expert in testing cells and tissue to find disease.

physical therapist
An expert in helping people move better.

plastic surgeon
A doctor who's an expert in operations to improve function and appearance.

positron emission tomography (PET)
A test that uses radioactive material to see the shape and function of body parts.

primary tumor
The first and main tumor.

progression
The growth or spread of cancer after being tested or treated.

radiation oncologist
A doctor who's an expert in treating cancer with radiation.

radiation therapy
A local treatment that uses high-energy rays or related approaches to kill cancer cells.

radiofrequency ablation (RFA)
A treatment that destroys cancer cells by heating them with high-energy radio waves.

radiologist
A doctor who's an expert in interpreting imaging tests.

recurrence
The return or worsening of cancer after a period of improvement.

resectable
Cancer that can be completely removed with surgery.

retroperitoneum
The body space in front of the spine in the lower trunk.

rhabdomyosarcoma (RMS)
A type of soft tissue sarcoma that starts in the cells that normally develop into skeletal muscle.

sarcoma
A cancer of bone or soft tissue cells.

stereotactic body radiation therapy (SBRT)
Radiation therapy given in higher doses to smaller areas over 1 to 5 sessions of treatment.

supportive care
Health care that includes symptom relief but not cancer treatment. Also called palliative care.

surgical margin
The normal-looking tissue around the edge of a tumor removed during surgery.

targeted therapy
A drug treatment that impedes the growth process specific to cancer cells.

thoracic surgeon
A doctor who's an expert in operating on organs inside the chest.

ultrasound
A test that uses sound waves to take pictures of the insides of the body.

unresectable
Cancer that can't be removed with surgery.

x-ray
A non-invasive imaging test that uses a small amount of radiation. Also called a plain radiograph.

NCCN Contributors

This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Soft Tissue Sarcoma, Version 1.2024. It was adapted, reviewed, and published with help from the following people:

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NCCN Cancer Centers

Abramson Cancer Center
at the University of Pennsylvania
Philadelphia, Pennsylvania
800.789.7366 • pennmedicine.org/cancer

Case Comprehensive Cancer Center/
University Hospitals Seidman Cancer Center and
Cleveland Clinic Taussig Cancer Institute
Cleveland, Ohio
UH Seidman Cancer Center
800.641.2422 • uhhospitals.org/services/cancer-services
CC Taussig Cancer Institute
866.223.8100 • my.clevelandclinic.org/departments/cancer
Case CCC
216.844.8797 • case.edu/cancer

City of Hope National Medical Center
Duarte, California
800.826.4673 • cityofhope.org

Dana-Farber/Brigham and Women's Cancer Center |
Mass General Cancer Center
Boston, Massachusetts
877.442.3324 • youhaveus.org
617.726.5130 • massgeneral.org/cancer-center

Duke Cancer Institute
Durham, North Carolina
888.275.3853 • dukecancerinstitute.org

Fox Chase Cancer Center
Philadelphia, Pennsylvania
888.369.2427 • foxchase.org

Fred & Pamela Buffett Cancer Center
Omaha, Nebraska
402.559.5600 • unmc.edu/cancercenter

Fred Hutchinson Cancer Center
Seattle, Washington
206.667.5000 • fredhutch.org

Huntsman Cancer Institute at the University of Utah
Salt Lake City, Utah
800.824.2073 • healthcare.utah.edu/huntsmancancerinstitute

Indiana University Melvin and Bren Simon
Comprehensive Cancer Center
Indianapolis, Indiana
888.600.4822 • www.cancer.iu.edu

Mayo Clinic Comprehensive Cancer Center
Phoenix/Scottsdale, Arizona
Jacksonville, Florida
Rochester, Minnesota
480.301.8000 • Arizona
904.953.0853 • Florida
507.538.3270 • Minnesota
mayoclinic.org/cancercenter

Memorial Sloan Kettering Cancer Center
New York, New York
800.525.2225 • mskcc.org

Moffitt Cancer Center
Tampa, Florida
888.663.3488 • moffitt.org

O'Neal Comprehensive Cancer Center at UAB
Birmingham, Alabama
800.822.0933 • uab.edu/onealcancercenter

Robert H. Lurie Comprehensive Cancer Center
of Northwestern University
Chicago, Illinois
866.587.4322 • cancer.northwestern.edu

Roswell Park Comprehensive Cancer Center
Buffalo, New York
877.275.7724 • roswellpark.org

Siteman Cancer Center at Barnes-Jewish Hospital
and Washington University School of Medicine
St. Louis, Missouri
800.600.3606 • siteman.wustl.edu

St. Jude Children's Research Hospital/
The University of Tennessee Health Science Center
Memphis, Tennessee
866.278.5833 • stjude.org
901.448.5500 • uthsc.edu

Stanford Cancer Institute
Stanford, California
877.668.7535 • cancer.stanford.edu

The Ohio State University Comprehensive Cancer Center -
James Cancer Hospital and Solove Research Institute
Columbus, Ohio
800.293.5066 • cancer.osu.edu

The Sidney Kimmel Comprehensive
Cancer Center at Johns Hopkins
Baltimore, Maryland
410.955.8964
www.hopkinskimmelcancercenter.org

The UChicago Medicine Comprehensive Cancer Center
Chicago, Illinois
773.702.1000 • uchicagomedicine.org/cancer

The University of Texas MD Anderson Cancer Center
Houston, Texas
844.269.5922 • mdanderson.org

UC Davis Comprehensive Cancer Center
Sacramento, California
916.734.5959 • 800.770.9261
health.ucdavis.edu/cancer

UC San Diego Moores Cancer Center
La Jolla, California
858.822.6100 • cancer.ucsd.edu

UCLA Jonsson Comprehensive Cancer Center
Los Angeles, California
310.825.5268 • uclahealth.org/cancer

UCSF Helen Diller Family
Comprehensive Cancer Center
San Francisco, California
800.689.8273 • cancer.ucsf.edu

University of Colorado Cancer Center
Aurora, Colorado
720.848.0300 • coloradocancercenter.org

University of Michigan Rogel Cancer Center
Ann Arbor, Michigan
800.865.1125 • rogelcancercenter.org

University of Wisconsin Carbone Cancer Center
Madison, Wisconsin
608.265.1700 • uwhealth.org/cancer

UT Southwestern Simmons
Comprehensive Cancer Center
Dallas, Texas
214.648.3111 • utsouthwestern.edu/simmons

Vanderbilt-Ingram Cancer Center
Nashville, Tennessee
877.936.8422 • vicc.org

Yale Cancer Center/Smilow Cancer Hospital
New Haven, Connecticut
855.4.SMILOW • yalecancercenter.org

Notes

Index

DR SHIVAM SHINGLA

BSES MG Hospital (Andheri):
9 am to 10 am (Monday to Friday)
Nanavati Max Hospital (Vile Parle):
10 am to 12 pm (Monday to Saturday)
S. L. Raheja Hospital (Mahim):
12 pm to 4 pm (Monday to Saturday)
Suvarna Hospital (Borivali):
5 pm to 6 pm (Monday and Friday)
Sushrut Hospital (Chembur):
By appointment
Hinduja Hospital (Khar): By appointment
Galaxy Healthcare (Borivali): By appointment

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#Reference From NCCN Guidelines