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Soft Tissue Sarcoma

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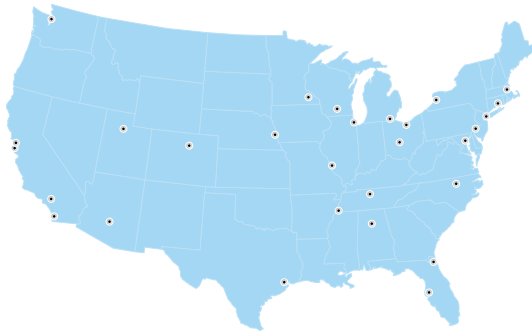
LEARNING that you have cancer can be overwhelming.

The goal of this book is to help you get the best care. It explains which cancer tests and treatments are recommended by experts in soft tissue sarcoma among adults.

The National Comprehensive Cancer Network® (NCCN®) is a not-for-profit alliance of 27 of the world's leading cancer centers. Experts from NCCN have written treatment guidelines for doctors who treat soft tissue sarcoma. These treatment guidelines suggest what the best practice is for cancer care. The information in this patient book is based on the guidelines written for doctors.

This book focuses on the treatment of soft tissue sarcoma among adults. Key points of this book are summarized in the related [NCCN Quick Guide](#).™ NCCN also offers patient resources on lung cancer, melanoma, and many other cancer types. Visit [NCCN.org/patients](https://www.nccn.org/patients) for the full library of patient books, summaries, and other patient and caregiver resources.

About



These patient guidelines for cancer care are produced by the National Comprehensive Cancer Network® (NCCN®).

The mission of NCCN is to improve cancer care so people can live better lives. At the core of NCCN are the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®). NCCN Guidelines® contain information to help health care workers plan the best cancer care. They list options for cancer care that are most likely to have the best results. The NCCN Guidelines for Patients® present the information from the NCCN Guidelines in an easy-to-learn format.

Panels of experts create the NCCN Guidelines. Most of the experts are from NCCN Member Institutions. Their areas of expertise are diverse. Many panels also include a patient advocate. Recommendations in the NCCN Guidelines are based on clinical trials and the experience of the panelists. The NCCN Guidelines are updated at least once a year. When funded, the patient books are updated to reflect the most recent version of the NCCN Guidelines for doctors.

For more information about the NCCN Guidelines, visit NCCN.org/clinical.asp.

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NCCN Foundation was founded by NCCN to raise funds for patient education based on the NCCN Guidelines. NCCN Foundation offers guidance to people with cancer and their caregivers at every step of their cancer journey. This is done by sharing key information from leading cancer experts. This information can be found in a library of NCCN Guidelines for Patients® and other patient education resources. NCCN Foundation is also committed to advancing cancer treatment by funding the nation's promising doctors at the center of cancer research, education, and progress of cancer therapies.

For more information about NCCN Foundation, visit NCCNFoundation.org.

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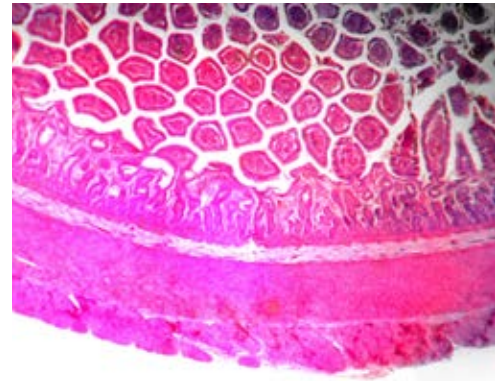
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SARC (Sarcoma Alliance for Research through Collaboration)

As a non-profit organization dedicated to the prevention, treatment, and cure of sarcomas, SARC understands the importance of empowering patients and caregivers with recommendations on the best care available. SARC strongly endorses the NCCN Guidelines for Patients in providing this vital resource for making informed decisions about their care. sarctrials.org



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Who should read this book?

This book is about treatment for soft tissue sarcoma in adults. Patients and those who support them—caregivers, family, and friends—may find this book helpful. It may help you discuss and decide with doctors what care is best.

Are the book chapters in a certain order?

Yes, information in early chapters explains treatment options found in later chapters. Starting with **Part 1** may be helpful for many people. It explains what sarcoma is. **Part 2** explains the tests doctors use to diagnose (confirm) a soft tissue sarcoma. **Part 3** describes the types of treatments that may be used.

The next 4 chapters offer a treatment guide for soft tissue sarcomas.

- **Part 4** shares treatment options for sarcomas in the arms, legs, trunk wall, head, or neck.
- **Part 5** covers sarcomas in the space in front of the lower spine (retroperitoneum), belly area (abdomen), and between the hip bones (pelvis).
- **Part 6** is a treatment guide for GISTs (**g**astrointestinal **s**tromal **t**umors).
- **Part 7** discusses options for desmoid tumors, and offers options for rhabdomyosarcoma.

Part 8 is the last chapter of the guideline. It offers some helpful tips for making treatment decisions. You can also get sample questions to ask your doctors. Visit the websites in this section for more on sarcoma.

Does this book include all options?

This book includes information for many situations. However, it doesn't address treatment for bone sarcomas (for example, Ewing sarcoma or osteogenic sarcoma) and desmoplastic small round cell tumors.

Your treatment team can help you with the options found in this guide. They can point out which sections apply to you. They can also give you more information. As you read through this book, you may find it helpful to make a list of questions to ask your doctors.

NCCN experts base the recommendations in this book on science and experience. However, these recommendations may not be right for your situation. Your doctors may suggest other tests and treatments based on your health and other factors. If other recommendations are given, feel free to ask your treatment team questions.

Help! What do the words mean?

In this book, many medical words are included. These are words you will likely hear from your treatment team. Most of these words may be new to you, and it may be a lot to learn.

Don't be discouraged as you read. Keep reading and review the information. Feel free to ask your treatment team to explain a word or phrase that you don't understand.

Words that you may not know are defined in the text or in the *Dictionary*. *Acronyms* are also defined when first used and in the Glossary. One example is FAP for **f**amilial **a**denomatous **p**olyposis.

1

Sarcoma basics

9 Soft tissue sarcoma

10 About cancer

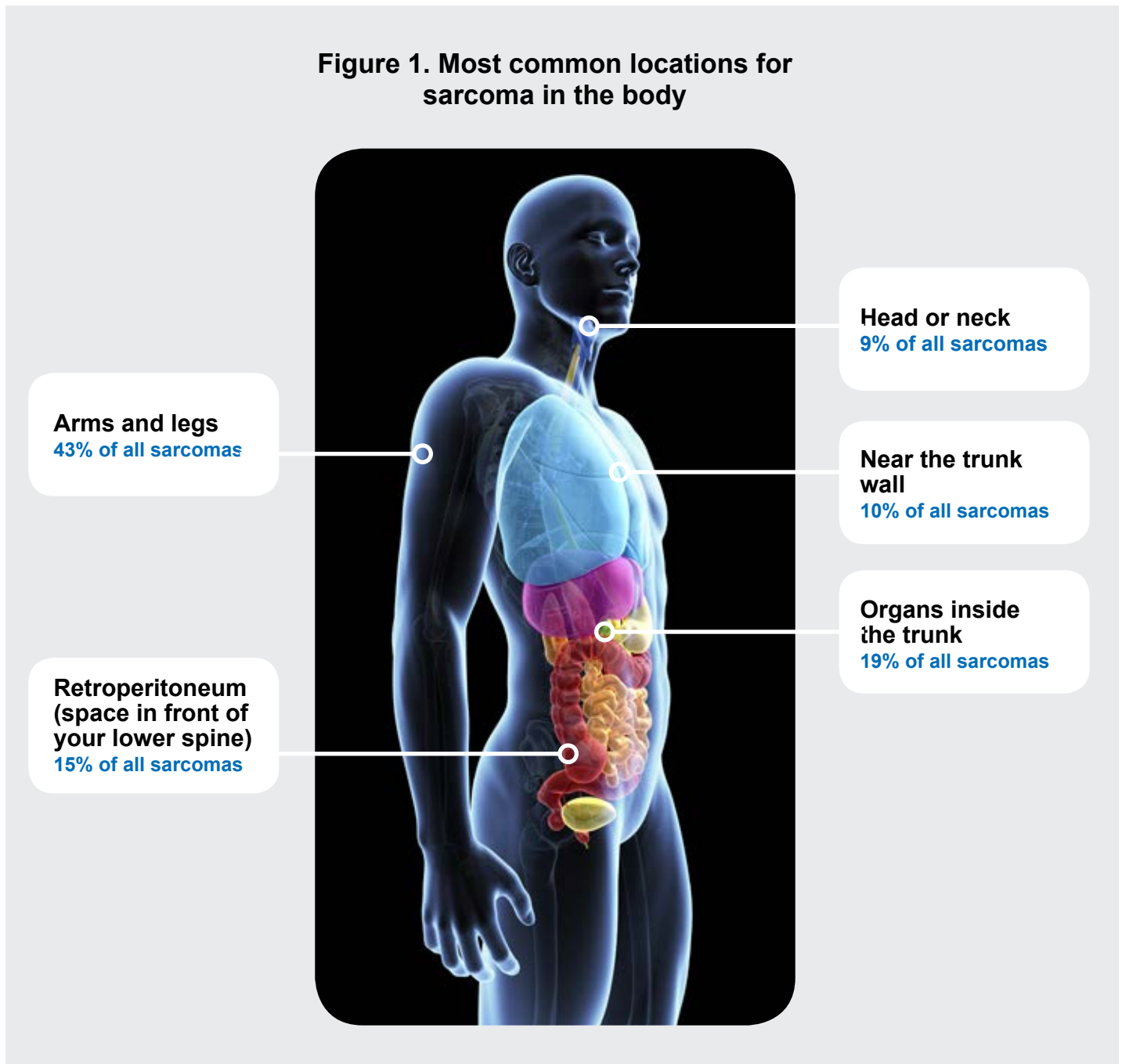
10 Review



Learning that you or a loved one have or may have cancer can be overwhelming. Part 1 describes sarcomas, and, most importantly for this guideline, explains a soft tissue sarcoma.

This chapter explains how this cancer starts in the body. It may also help you better understand this disease and talk with your doctor.

Figure 1. Most common locations for sarcoma in the body



Soft tissue sarcoma

Sarcomas are a large but rare group of cancers. Sarcomas start in cells that make up bones or soft tissue. Soft tissue supports, connects, and surrounds parts of your body. Soft tissue includes fat, muscles, nerves, tendons, and blood and lymph vessels. Soft tissue sarcomas are more common than bone sarcomas. Bone sarcomas, such as osteosarcomas, are often diagnosed in children.

There are over 50 types of soft tissue sarcoma. These types differ by the type of soft tissue. They also differ by the type and shape of cells, and how much the cells look like normal cells. A list of soft tissue sarcomas are listed on page 74. The most common types are:

- Undifferentiated pleomorphic sarcoma
- GISTs (gastrointestinal stromal tumors)
- Liposarcoma
- Leiomyosarcoma
- Synovial sarcoma
- Malignant peripheral nerve sheath tumors

Soft tissue sarcomas can occur anywhere in the body. The most common place is in the arms and legs (limbs). **See Figure 1.** About 43 out of 100 (43%) sarcomas occur in the limbs. The second most common site is in the abdomen, such as the stomach and intestines. About 19% of sarcomas occur here. Next, about 15% of sarcomas occur in a space in front of your lower spine called the retroperitoneum. About 10% of sarcomas occur in the trunk, and 9% occur in the head or neck. The trunk is also called the torso. It is the main part of the body not including the head, neck, or limbs.

This patient guideline will focus on:

- Soft tissue sarcomas in the limbs, outer trunk, head, or neck
- Retroperitoneal or intra-abdominal soft tissue sarcomas (inner trunk)
- GISTs
- Desmoid tumors (aggressive fibromatoses)
- Rhabdomyosarcoma



NCCN experts recommend that people with soft tissue sarcomas seek care with an experienced team of doctors. These doctors should specialize in the treatment of sarcomas.

It is helpful to discuss the treatment options available to you. Ask your treatment team questions about your care plan.

About cancer

Knowing more about how cancer starts may help you understand how a sarcoma forms in the body. Sarcoma is a type of cancer. Cancer is a disease that starts in the cells of your body. Cells are the building blocks of tissue in the body. The human body contains trillions of cells. DNA (deoxyribonucleic acid) is found within each cell and controls the cells, instructing them on what to do. The coded instructions for your cells found in DNA are called genes.

Normal cells grow and divide and repeat the process over and over again. Normal genes tell cells that they are supposed to die when they become old or damaged. If they don't die, and new cells start to form, this growth can get out of control. Abnormal out-of-control growth can be caused by a problem with the genes, and can lead to the growth of a solid mass of cells called a tumor. Some tumors are malignant (cancer). Some tumors are benign (not cancer). Solid tumors can grow anywhere in the body and can affect the way the body works.

Review

- ▶ Sarcomas are a large but rare group of cancers.
- ▶ Sarcomas start in cells that make up bones or soft tissue.
- ▶ Soft tissue includes fat, muscles, nerves, tendons, and blood and lymph vessels.

2

Testing for soft tissue sarcoma

12 Medical history & physical exam

12 Blood tests

13 Imaging tests

15 Genetic assessment

16 Biopsy

17 Review



Part 2 focuses on medical tests used for soft tissue sarcoma. These tests help diagnose (confirm) cancer. Some of the tests may continue during and after treatment. These tests help your doctor make a treatment plan.

Medical history & physical exam

Two basic tools of diagnosis include a medical history and physical exam. Your doctor will ask about your medical history, which should include everything that has ever happened to you, related to your health. He or she will also do an exam of your body.

Your doctor may ask you about:

- ▶ Health events in your life including surgeries, accidents, and past illnesses
- ▶ Recent sickness
- ▶ New symptoms (especially pain, bleeding, weight changes, cough, or breathing difficulties)
- ▶ Medications you are taking now and those you may be allergic to (It is helpful to keep a list of your medications. Include any supplements and over-the-counter medicines you take.)
- ▶ Family history of disease such as cancer, heart disease, or diabetes

When your doctor checks your body for signs of disease, it is called a physical exam. Doctors often perform a physical exam along with taking a medical history.

Your doctor may:

- ▶ Check your eyes, ears, nose, mouth, and neck
- ▶ Listen to your lungs, heart, and belly (abdomen)
- ▶ Feel or use pressure to see if organs are of normal size, soft or hard, or cause pain when touched

Blood tests

Blood tests are not used to diagnose a soft tissue sarcoma. Doctors test blood to look for signs of disease and assess your general health. They may be used to assess the response to drugs that are being used for treatment. Abnormal levels of certain chemicals in the blood may be a sign that the cancer has spread to distant parts of the body.

Complete blood count with differential

A CBC (complete blood count) measures the number of blood cells in a blood sample. It includes numbers of white blood cells, red blood cells, and platelets. Your blood counts may be low or high because of cancer or another health problem. It is an essential test that gives a picture of your overall health.

There are several types of white blood cells in your body. A white cell differential counts the number of each type of cell. It also checks if the counts are in balance with each other. This test can help your doctor learn the cause of an abnormal white blood cell count. It is also used to stage the cancer and check if treatment is working.

Comprehensive metabolic panel

Chemicals in your blood come from your liver, bone, and other organs. A comprehensive metabolic panel often includes tests for up to 14 chemicals. The tests show if the levels of chemicals are too low or high. Abnormal levels can be caused by cancer or other health problems.

Imaging tests

Imaging tests are used to take pictures (images) of the inside of your body. Imaging can be used to see if there is cancer in the body. Your doctor will want to check the primary tumor, or original site of the tumor. CT (computed tomography) and MRI (magnetic resonance imaging) scans are the recommended imaging tests for soft tissue sarcomas.

What to know about imaging tests:

- Imaging tests can see if the cancer is in more than one part of the body.
- They continue during treatment to see how the disease is responding.
- They can be used after treatment to check for signs of disease (recurrence).
- You may not learn of the results of your imaging tests for a few days since a radiologist needs to see the pictures.
- A radiologist is a doctor who is an expert in reading the pictures from imaging tests.

CT scan

A CT scan (**Figure 2**) uses x-rays to take pictures of the inside of the body. It takes many x-rays of the same body part from different angles. All the x-ray pictures are combined to make one detailed picture.

Figure 2. CT scan machine

A CT scan machine is large and has a tunnel in the middle. During the scan, you will need to lie face up on a table that moves through the tunnel. The scanner will rotate an x-ray beam around you to take pictures from many angles. A computer will combine all the x-ray pictures into one detailed picture.



A CT scan of your chest, abdomen, and/or pelvis may be one of the tests to look for cancer.

Before the CT scan, you may be given a contrast dye to make the pictures clearer. You may drink the dye, have it injected into your vein, or both. It may cause you to feel flushed or get hives. Rarely, serious allergic reactions occur. Tell your doctors if you have had bad reactions in the past.

MRI scan

An MRI scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays. This type of scan is good at showing the spine and soft tissues like the brain. An MRI scan can also be used to assess the abdomen and pelvis.

An MRI may be used as an initial test, to check treatment results, and to check for the spread of cancer to other parts of the body. Getting an MRI scan is similar to getting a CT scan, but MRI scans take longer to complete.

For the scan, you will need to lie on a table that moves through a large tunnel in the scanning machine. The scan may cause your body to feel a bit warm. Like a CT scan, a contrast dye may be used to make the pictures clearer.

PET scan

A PET (**p**ositron **e**mission **t**omography) scan shows how active your cells are by showing how fast they use up a simple form of sugar called glucose. To create pictures, a sugar radiotracer first needs to be put into your body by an injection into a vein. The radiotracer emits a small amount of energy that is detected by the machine that takes pictures. Active cancer cells use sugar faster than normal cells. Thus, cancer cells look brighter in the pictures.

PET is very good at showing small groups of cancer cells. This test may also be useful for showing if sarcoma has spread. Sometimes, PET is combined with CT—called a PET/CT (**p**ositron **e**mission **t**omography/**c**omputed **t**omography) scan.

X-ray

An x-ray uses small amounts of radiation to make pictures of organs and tissues inside the body. A tumor changes the way radiation is absorbed and will show up on the x-ray picture. A chest x-ray may be used with other initial tests when sarcoma is first suspected or found. It may also be done after treatment to check treatment results.

Ultrasound

An ultrasound is a test that uses sound waves to form pictures of the inside of the body. Ultrasound is good at showing small areas of cancer that are superficial (on the surface of the body). Ultrasounds are generally painless.

Angiogram

An angiogram is a test that uses an x-ray to check the blood vessels and the flow of blood to detect any blockage or leakage. This type of test uses contrast put into a catheter (thin, flexible tube) to assess the blood vessels. A doctor will insert the catheter into an artery, so you will be asked to lie still during the test. You may be sore after the test and be asked to rest at home.

Genetic assessment

NCCN experts suggest genetic counseling for people with a known or possible personal or family history of hereditary cancer syndromes. Hereditary means it is passed down through families (inherited). Some syndromes related to a soft tissue sarcoma include:

- Li-Fraumeni syndrome
- FAP (familial adenomatous polyposis)
- Gardner's syndrome
- Carney-Stratakis syndrome

About the syndromes

Genes provide the instructions cells use to make new cells and control how cells behave. An abnormal change in these instructions—called a gene mutation—can cause cells to grow and divide out of control. Families with Li-Fraumeni syndrome have a gene mutation (error) in the *TP53* gene. *TP53* is the gene that 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.

People with FAP have an *APC* (adenomatous polyposis coli) gene mutation. This syndrome causes colon polyps (usually many) that may become colon cancer. A polyp is a growth from the inner wall of the digestive tract. FAP has a related syndrome called Gardner's syndrome and both are associated with desmoid tumors (see Part 6 on page 46).

Carney-Stratakis syndrome puts someone at risk for GIST and paragliomas. GIST is a soft tissue sarcoma usually found in the stomach or small intestine, and a paraglioma is a benign tumor (not cancer) found in the head, neck, or upper body. These patients have mutations in a family of *SDH* genes.

GISTs with an *SDH* mutation usually occur in the stomach of younger people, often spread, and may be slow-growing. See Part 5 for more information on GIST.

Other gene mutations may also be associated with certain types of sarcoma. A genetic counselor is an expert who has special training in genetic diseases. Genetic counselors can help people understand the risk of developing sarcoma or other cancers because of genetic mutations.

Ask your treatment team about genetic diseases and cancer. To learn more about cancer risk and prevention, and treatment of cancer by type, see the NCCN Guidelines®, available for free at www.nccn.org.

- For more on Li-Fraumeni syndrome, see the *NCCN Guidelines for Genetic/Familial High-Risk Assessment: Breast and Ovarian*.
- For more on FAP, see the *NCCN Guidelines for Genetic/Familial High-Risk Assessment: Colorectal*.
- For more on Carney-Stratakis syndrome and hereditary cancer syndromes for sarcoma, see the *NCCN Guidelines for Soft Tissue Sarcoma*.

Biopsy

Tissue or fluid may be removed from your body and tested to diagnose (confirm) cancer. A biopsy is a procedure that removes samples of fluid or tissue. Sometimes a sample of tissue from the biopsy does not have enough cells to check for cancer. The tissue could also be abnormal but not cancer. If this happens, you may have another biopsy.

After imaging tests, a small sample of tissue from the primary tumor may be removed by a type of biopsy listed below:

- ▶ Core needle biopsy removes tissue samples with a hollow needle. This is usually the preferred biopsy for sarcoma.
- ▶ Incisional biopsy removes a small amount of tissue.
- ▶ FNA (**f**ine-**n**eedle **a**spiration) biopsy uses a thin needle to take a sample of tissue. An ultrasound may guide the FNA biopsy.

Other image-guided needle biopsies may be needed for tumors in the chest, abdomen, pelvis, or deep areas. The biopsy samples will be sent to a pathologist. A pathologist is a doctor who is an expert in examining cells to find disease.

Test results and next steps

Once your doctors review your test results, they may talk to you about your next steps of care. Talking with your doctor about your diagnosis can help with treatment planning. Shared decision-making is a process in which you and your doctors decide on the type(s) of treatment together. Shared-decision making is an important part of your treatment plan.

Shared decision-making also involves thinking about the things that will change in your everyday life. For example, you may think about:

- ▶ How treatment will affect your health and normal activities
- ▶ If you might stay in the hospital for days or need in-home care
- ▶ Whether or not you may need daily help from others

You may want to make a list of questions for your treatment team. It may also be helpful to talk with your family or friends as you make decisions about treatment.

Review

- ▶ Cancer tests are used to find cancer, plan treatment, and check how well treatment is working.
- ▶ Your health history and a body exam inform your doctor about your health.
- ▶ Blood tests check for signs of disease.
- ▶ Imaging tests that take pictures of the inside of your body may show cancer.
- ▶ A biopsy removes tissue or fluid from your body to diagnose (confirm) cancer.
- ▶ Shared decision-making is a process in which you and your doctors decide on the type(s) of treatment together.

What to know...

- ✓ Your doctors will order tests and schedule visits to talk about your care plan.
- ✓ It is helpful to keep track of your test results at all times.
- ✓ Ask your doctors questions about the results.

3

Overview of cancer treatments

19 Treatment planning

20 Surgery

20 Radiation therapy

22 Chemotherapy

22 Targeted therapy

23 Immunotherapy

26 Clinical trials

27 Review



Part 3 describes the main treatments for sarcoma. Knowing what a treatment is will help you understand your treatment options listed in each *Treatment guide* chapter. There is more than one treatment for sarcoma. Not every person with sarcoma will receive every treatment listed in this chapter.

Treatment planning

Treatment team

Treatment of sarcoma takes a team of experts who have experience with this cancer. This includes oncologists and other medical or health care staff. The word "oncologist" means cancer doctor.

If you have sarcoma, it is important that the experts meet before your treatment starts. They will meet to create the best treatment plan for you. Your treatment team will also meet while you are going through treatment. After treatment is complete, they will discuss the results and plan your next steps of care. Your team of experts may include:

In most cases:

- **Pathologist**—an expert in testing cells and tissue to find and define the type of disease
- **Radiologist**—an expert in imaging tests
- **Oncology surgeon**—an expert in operations that remove cancer
- **Medical oncologist**—an expert in cancer drugs

- **Radiation oncologist**—an expert in radiation treatment
- **Nurse**—an expert trained to care for patients

In some cases:

- **Thoracic surgeon**—an expert in operations within the chest
- **Gastroenterologist**—an expert in digestive diseases
- **Plastic surgeon**—an expert in operations to improve function and appearance
- **Social worker**—an expert in meeting social and emotional needs
- **Occupational therapist**—an expert in helping people with the tasks of daily living
- **Physical therapist**—an expert in helping people move with greater comfort and ease
- **Nutritionist**—an expert in healthy foods and drinks
- **Genetic counselor**—an expert in explaining and testing for hereditary diseases



Types of treatment options

You will learn more about local and systemic treatment options in this chapter. Local treatments are used to treat a focused area of cancer. Surgery and radiation therapy are common local treatments. Ablation and embolization are also local treatments. They are sometimes used for small tumors that aren't near where the cancer started. An example is small tumors in the liver.

Systemic treatments are able to treat cancer cells throughout the body. This type of treatment is an option for many people with sarcoma. It comes in the form of drug treatment that includes chemotherapy, targeted therapy, and immunotherapy.

Surgery

Surgery is a primary treatment for sarcoma. Primary treatment is the main treatment used to rid the body of cancer. The goal of surgery is to remove all the cancer. Surgery may also be used to reduce symptoms caused by the cancer or extend life. This is called palliative or supportive care.

The method and extent of surgery for sarcoma varies. Both depend on where the sarcoma is and the size of the tumor. Ask your treatment team for details of how the tumor will be removed.

Your surgeon will try to obtain a cancer-free surgical margin. A surgical margin is a ring of normal-looking tissue around the tumor. Sometimes surgeons can't safely remove the sarcoma with a cancer-free margin. If this is expected, your surgeon will place clips in your body after removing the tumor. The clips help doctors know where to give you radiation therapy. Sometimes, a second surgery can be done instead of removing the remaining cancer.

Your doctor will also try to remove as little tissue as possible. This is done so you can use your limbs or other body parts after surgery. For some people, their limb may need to be removed (amputated) in order to remove all the cancer.

Possible side effects of surgery

Side effects are unhealthy or unpleasant physical or emotional responses to treatment. You may experience side effects from the anesthesia or surgery. Often, general anesthesia is used for surgery. General anesthesia uses drugs that put you into a deep sleep-like state so you won't feel pain. Ask your treatment team for a full list of possible side effects of the surgery you will have.

Radiation therapy

Radiation therapy uses high-energy rays to treat cancer. The rays damage DNA. DNA is a chain of chemicals in cells that contains genes. Radiation therapy either kills the cancer cells or stops new cancer cells from being made. It has many uses for sarcomas.

Radiation may be given as:

- ▶ Neoadjuvant therapy before surgery to shrink the tumor
- ▶ Primary (first) treatment for sarcoma
- ▶ IORT (intraoperative radiation therapy) during surgery
- ▶ Adjuvant treatment, after surgery is complete
- ▶ A full dose of radiation or, if receiving a "boost," less than a full dose

There are different ways to give radiation. Which method you get depends on the type of sarcoma and the purpose of radiation therapy. Some methods are discussed next. You may feel side effects from radiation, although not everyone does. Ask your treatment team for a full list of side effects.

External beam radiation therapy

Radiation is often given using a machine outside the body. This method is called EBRT (**e**xternal **b**eam **r**adiation **t**herapy). To receive EBRT, you first must have a simulation session. For simulation, imaging scans are used to help target the tumor with radiation.

Using the scans, your treatment team will plan the best radiation dose, number and orientation of radiation beams, and number of treatment sessions. Radiation beams are directed at the tumor with help from images taken prior to each treatment, ink marks on the skin, and/or marker seeds in the tumor.

During treatment, you will lie on a table in the same position as you did for simulation. Devices may be used to keep you from moving so that the radiation targets the tumor in the same place every day of treatment. You will be alone while the technician operates the machine from a nearby room. He or she will be able to see, hear, and speak with you at all times. As treatment is given, you may hear noises.

There are multiple types of EBRT to treat sarcoma:

- 3D-CRT (**t**hree-**d**imensional **c**onformal **r**adiation **t**herapy) uses beams matched to the shape of the tumor
- IMRT (**i**ntensity-**m**odulated **r**adiation **t**herapy) uses small beams of different strengths

- SBRT (**s**tereotactic **b**ody **r**adiation **t**herapy) involves high-dose radiation within one or a few sessions

Brachytherapy

Brachytherapy involves placing small radioactive objects next to cancer cells. The objects are inserted using small tubes (catheters) that were placed during surgery. Brachytherapy can be given either as LDR (**l**ow-**d**ose **r**ate) or HDR (**h**igh-**d**ose **r**ate). The dose rate has to do with the intensity level of the treatment. Your doctor can discuss this treatment with you if it is recommended.

Ablation

Ablation destroys small tumors with little harm to nearby tissue. There is more than one way to ablate a tumor. Cryoablation kills cancer cells by freezing them with liquid nitrogen. Radiofrequency ablation kills cancer cells with high-energy radio waves. A probe placed into the tumor emits the waves. The probe will be guided into place by ultrasound or CT scans and will be removed when treatment is done.

Embolization

Embolization treats tumors by cutting off their blood supply. A catheter will be inserted into an artery and guided to the tumor. Once in place, beads will be inserted to block the blood flow. With chemoembolization, the beads are coated with chemotherapy. Radioembolization uses small radioactive beads.

Drugs can also be given to treat cancer throughout the body. This is called systemic therapy. Doctors use systemic drugs to treat cancer cells that may have spread beyond the first site of cancer. The types of drugs used for sarcoma include chemotherapy, targeted therapy, and immunotherapy. See **Guide 1** on pages 24 and 25 for a list of drugs used for treating sarcoma.

Chemotherapy

Chemotherapy is the use of drugs to destroy abnormal cells in the body. But, the drugs can also affect normal cells. Many people refer to this treatment as “chemo.”

Different types of chemotherapy drugs work in different ways to kill abnormal cells or stop new ones from being made. Thus, more than one drug may be used. When only one drug is used, it's called a single agent. A combination regimen is the use of two or more chemotherapy drugs.

Some chemotherapy drugs are liquids that are injected into a vein or under the skin with a needle. Other chemotherapy drugs may be given as a pill that is swallowed.

Chemotherapy is given in cycles of treatment days followed by days of rest. This allows the body to recover before the next cycle. 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.

Chemotherapy and radiation given together is called chemoradiation. Sometimes these treatments are given at the same time, while other times they are staggered (for example, chemotherapy is given, then radiation, and then more chemotherapy).

Possible side effects of chemotherapy

The side effects of chemotherapy can differ between people. Some people have many side effects. Others have few. Some side effects can be very serious while others can be unpleasant but not serious. Side effects of chemotherapy depend on the drug type, amount taken, length of treatment, and the person.

Some common side effects of chemotherapy include low blood cell counts, not feeling hungry, nausea, vomiting, diarrhea, hair loss, and mouth sores. Please ask your treatment team for a complete list of side effects.

Targeted therapy

Targeted therapy stops the action of molecules involved in the growth of cancer cells. Some targeted therapy drugs block the chemical signals that tell the sarcoma cells to grow. Other targeted therapy drugs block signals that cause new blood vessels to form. Other drugs target hormones.

Targeted therapy isn't used for every sarcoma. Ask your doctor if targeted therapy may help you. Also ask about side effects. Targeted therapy harms normal cells less than chemotherapy but still has side effects. Side effects differ between drugs. Most targeted therapies come in pill form but some need to be injected.

Possible side effects of targeted therapy

The side effects of targeted therapy depend on the drug and dose. Some of the side effects listed are caused by only one targeted drug. Other side effects are caused by many targeted drugs but differ in how likely they are to occur.

Some common side effects of targeted therapy drugs used for sarcoma are tiredness, joint pain, skin rash, muscle pain, swelling, headache, fever, nausea or vomiting, and diarrhea. These drugs may also cause low blood cell counts.

Not all side effects of targeted therapy drugs are listed here. Be sure to ask your treatment team for a complete list of side effects. If a side effect bothers you, let your treatment team know how you are feeling.

Immunotherapy

The immune system is the body's natural defense against infection and disease. The immune system includes many chemicals and proteins. These chemicals and proteins are made naturally in your body.

Immunotherapy increases the activity of your immune system. By doing so, it improves your body's ability to find and destroy cancer cells. Interferon alfa is an immunotherapy used for desmoid tumors.

Possible side effects of immunotherapy

The side effects of immunotherapy depend on the drug and dose. Some common side effects of interferon alfa are flu-like symptoms, nausea, vomiting, not feeling hungry, depression, hair thinning, and liver damage. Ask your doctor about the side effects of immunotherapy.

Guide 1. Drug treatment for sarcoma

Generic name	Brand name (sold as)	Type of drug
Chemotherapy		
Carboplatin	–	Chemotherapy
Cyclophosphamide	–	Chemotherapy
Dacarbazine	–	Chemotherapy
Dactinomycin	Cosmegen®	Chemotherapy
Docetaxel	Taxotere®	Chemotherapy
Doxorubicin hydrochloride	–	Chemotherapy
Doxorubicin hydrochloride, liposome injection	Doxil®	Chemotherapy
Epirubicin hydrochloride	Ellence®	Chemotherapy
Eribulin	Halaven®	Chemotherapy
Etoposide	–	Chemotherapy
Etoposide phosphate	Etopophos®	Chemotherapy
Gemcitabine hydrochloride	Gemzar®	Chemotherapy
Ifosfamide	–	Chemotherapy
Irinotecan hydrochloride	Camptosar®	Chemotherapy
Methotrexate	–	Chemotherapy
Methotrexate sodium	–	Chemotherapy
Paclitaxel	Taxol®	Chemotherapy
Topotecan	Hycamtin® Capsules	Chemotherapy
Topotecan hydrochloride	Hycamtin® Injection	Chemotherapy
Temozolomide	Temodar®	Chemotherapy
Trabectedin	Yondelis®	Chemotherapy
Vincristine sulfate	–	Chemotherapy
Vinblastine sulfate	–	Chemotherapy
Vinorelbine tartrate	Navelbine®	Chemotherapy

Guide 1 (continued). Drug treatment for sarcoma

Generic name	Brand name (sold as)	Type of drug
Targeted therapy		
Bevacizumab	Avastin®	Targeted therapy
Ceritinib	Zykadia®	Targeted therapy
Crizotinib	Xalkori®	Targeted therapy
Dasatinib	Sprycel®	Targeted therapy
Everolimus	Afinitor®	Targeted therapy
Imatinib mesylate	Gleevec®	Targeted therapy
Nilotinib hydrochloride monohydrate	Tasigna®	Targeted therapy
Palbociclib	Ibrance®	Targeted therapy
Pazopanib	Votrient®	Targeted therapy
Regorafenib	Stivarga®	Targeted therapy
Sirolimus	Rapamune®	Targeted therapy
Sorafenib tosylate	Nexavar®	Targeted therapy
Sunitinib malate	Sutent®	Targeted therapy
Temsirolimus	Torisel®	Targeted therapy

Immunotherapy and other types

Interferon alfa 2b, recombinant	Intron® A	Immunotherapy
Celecoxib	Celebrex®	NSAID (nonsteroidal anti-inflammatory drug)
Mesna	Mesnex® Injection Mesnex® Tablets	Cytoprotective agent (used in combination with ifosfamide)
Sulindac	Clinoril®	NSAID
Tamoxifen citrate	–	Nonsteroidal antiestrogen
Toremifene citrate	Fareston®	Nonsteroidal antiestrogen

Clinical trials

Clinical trials are research studies that people choose to take part in. Because of clinical trials, doctors learn how to prevent, diagnose, and treat diseases like sarcoma. Because of clinical trials, the tests and treatments in this book are now widely used to help people with sarcoma.

One of your treatment choices may be to join a clinical trial. NCCN experts strongly support clinical trials as a treatment option. Clinical trials are an important option for people with soft tissue sarcoma.

Complementary and alternative medicine



CAM (**c**omplementary and **a**lternative **m**edicine) is a group of treatments sometimes used by people with cancer. Many CAMs are being studied to see if they are truly helpful.

Complementary medicines are meant to be used alongside standard therapies, most often for relaxation, improving your health, or to prevent or reduce side effects.

Alternative medicine is treatment or techniques that are used instead of standard treatments such as chemotherapy or radiation. Some are sold as cures even though they haven't been proven to work in clinical trials.

Many cancer centers or local hospitals have complementary therapy programs that offer acupuncture, yoga, and other types of therapy.

It's important to tell your treatment team if you are using any complementary medicines, especially supplements, vitamins, or herbs. Some of these things can interfere with your cancer treatment. For more information about CAM, ask your doctor and visit the websites listed in Part 8.

Phases of a clinical trial

Clinical trials go through levels or phases of testing to find safe and helpful ways to manage sarcoma. These phases help move the research along to find out what works best for patients with cancer.

- **Phase I** looks at how much of a drug to give, the drug's side effects, and how often to give the treatment.
- **Phase II** tests for side effects and whether a drug works for a specific type of cancer.
- **Phase III** compares the new treatment (or new use of treatment) to what is commonly used.
- **Phase IV** follows late side effects and determines whether the treatment still works after a long period of time.

Taking part in a clinical trial

All clinical trials have a plan and are carefully led by a medical team. Patients in a clinical trial are often alike with their cancer type and general health. You can join a clinical trial when you meet certain terms (eligibility criteria).

If you decide to join a clinical trial, you will need to review and sign a paper called an informed consent form. This form describes the clinical trial in detail, including the risks and benefits. Even after you sign consent, you can stop taking part in a clinical trial at any time.

Some benefits of a clinical trial:

- You'll have access to the most current cancer care.
- You will be closely watched by your medical team.

- You may help other patients with cancer.

Some risks of a clinical trial:

- Like any test or treatment, there may be side effects.
- New tests or treatments may not work.
- You may have to visit the hospital more often for treatment and appointments.

Next steps

Ask your doctor or nurse if a clinical trial may be an option for you. There may be clinical trials where you're getting treatment or at other treatment centers nearby. You can also find clinical trials through the websites listed in Part 8.

Review

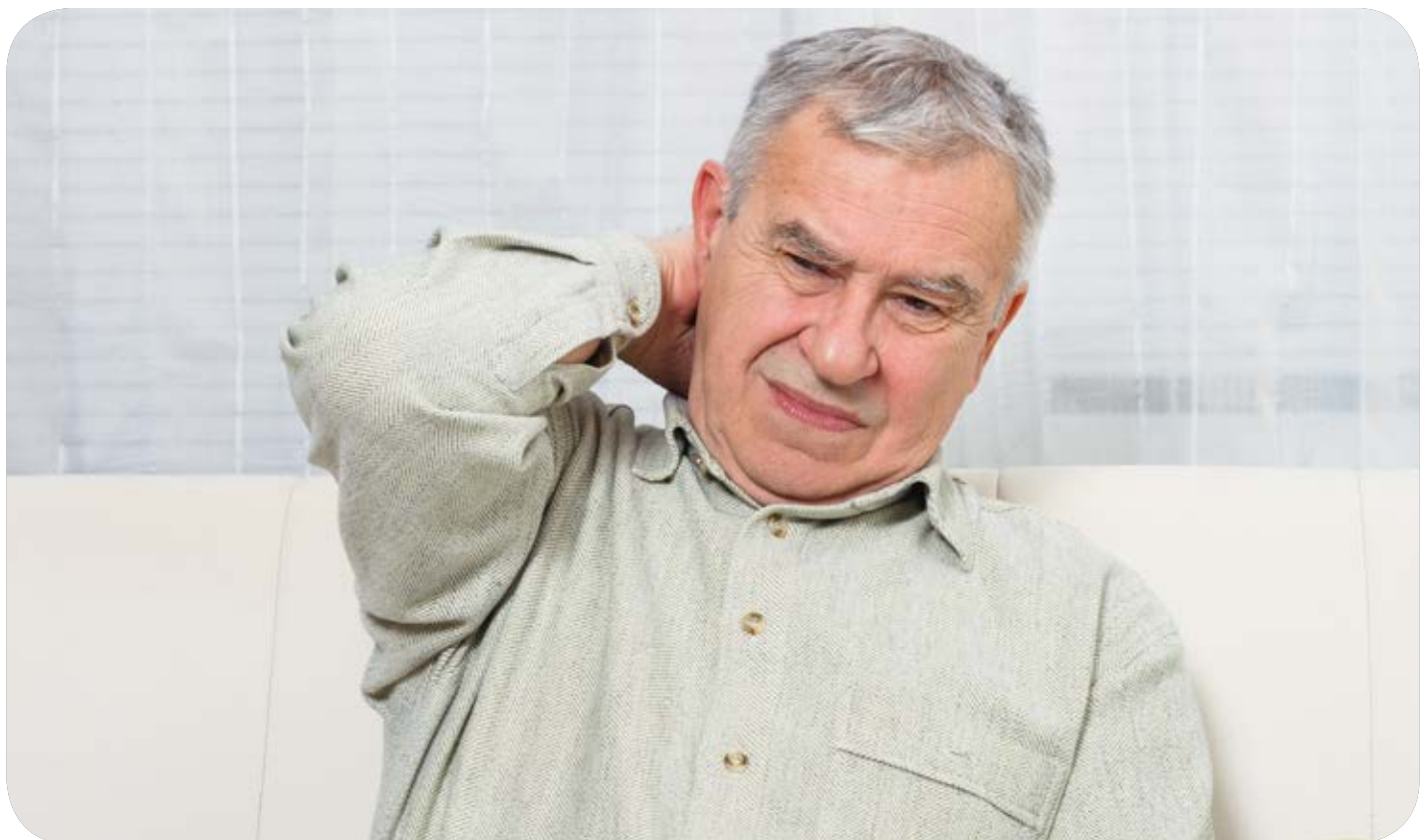
- Local treatments for soft tissue sarcoma include surgery, radiation therapy, ablation, and embolization.
- Systemic treatments are able to treat cancer cells throughout the body.
- Systemic treatments include drug treatments such as chemotherapy, targeted therapy, and immunotherapy.
- Clinical trials help doctors learn how to prevent, diagnose, and treat a disease like cancer.

4

Treatment guide

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

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30	Treatment by stage
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34	Stages II and III sarcoma
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39	Recurrence treatment
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Part 4 is a guide through the treatment options for sarcomas in the arms, legs, outer trunk, head, or neck. It starts by listing what is needed for treatment planning. Treatment options are then presented by cancer stage and if the cancer returns.

This information is taken from the treatment guidelines written by NCCN experts of sarcoma. These treatment guidelines list options for people with sarcoma in general. Thus, your doctors may suggest other treatment for you based on your health and personal wishes.



Helpful tips for reading the treatment guides in the next 4 chapters ➡

- ✓ The treatment guides (that look like charts) are numbered and list test or treatment options.
- ✓ It is helpful to read the guides from left to right, using the arrows to follow a path. The titles of the guides are important.
- ✓ The symbol \pm means with or without. This is used to show when a test or treatment is given "with or without" another test or treatment.
- ✓ Acronyms (a group of letters using the first letters of words or phrases) found in the treatment guides are defined on page 73.

Treatment planning

A team of experts who have experience with sarcoma should be managing your care. The team should create a plan designed for sarcoma in the limbs, outer trunk, head, or neck. In order to make a plan, certain tests will be done to find out more about your health. Your doctors will take a medical history, do an exam of your body, order imaging tests, and do a biopsy. [See Guide 2.](#)

Imaging tests of the primary tumor will be helpful to get a baseline (starting) measure of the cancer. This includes an MRI with or without a CT scan. Other imaging tests may be done to assess a certain part of the body. This is based on the type of soft tissue sarcoma, location in the body, and where the sarcoma may spread. For example, if the sarcoma may spread to the CNS (central nervous system) an MRI or CT scan of the brain and spinal cord may be suggested.

A biopsy should be done to confirm cancer and assess the specific cell type of sarcoma. A core needle biopsy is preferred. An incisional biopsy guided by an imaging test or an FNA biopsy may also be done in some cases. If your doctor wants to check for genetic abnormalities, you may meet with a genetics counselor. He or she will talk with you about your personal or family history of disease.

Treatment by stage

The treatment options for sarcomas in this chapter are listed by stage. Cancer staging is a rating by your doctors of the extent of the cancer. It is used to plan which treatments are best for you. The AJCC (American Joint Committee on Cancer) staging system is often used to stage sarcoma.

In this system, the letters T (tumor), N (node), and M (metastasis) describe an area of cancer growth.

Guide 2. Tests for soft tissue sarcoma in the limbs, outer trunk, head, or neck

Needed for most patients	May be needed for some patients
<ul style="list-style-type: none"> • Medical history and physical exam 	<ul style="list-style-type: none"> • PET/CT
<ul style="list-style-type: none"> • MRI ± CT scan of primary tumor 	<ul style="list-style-type: none"> • CT of abdomen and pelvis
<ul style="list-style-type: none"> • X-ray or CT of chest (preferred) 	<ul style="list-style-type: none"> • MRI of total spine
<ul style="list-style-type: none"> • Biopsy 	<ul style="list-style-type: none"> • Imaging of CNS
	<ul style="list-style-type: none"> • Angiogram and x-ray
	<ul style="list-style-type: none"> • Genetic assessment

The T, N, and M scores are combined to assign the cancer a stage.

- T score describes the growth of the primary tumor.
- N score describes spread of cancer growth to lymph nodes.
- M score tells if the cancer has spread to distant sites.

Another factor used in staging is the cancer grade. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. The letter G stands for the grade. GX means the grade can't be assessed, followed by G1, G2, and G3. G3 is the highest grade.

Other widely used grading systems include those by the FNCLCC (French Federation of Cancer Centers Sarcoma Group) and the NCI (National Cancer Institute). Each system uses a 3-part scoring system as listed below:

- How much the cancer cells look like normal cells (FNCLCC) or the specific cell type (NCI)
- How fast the cancer cells are making copies of themselves (FNCLCC) or tumor location (NCI)
- How much dead tissue is in the tumor (FNCLCC and NCI)

AJCC groups the TNM and grade into 4 stages— I, II, III, and IV. Stages I–IV are used for sarcomas in the trunk, limbs, retroperitoneum, and for GIST tumors.

Doctors use the staging and grading information of different types of sarcomas in many ways. Thus, staging for sarcoma may be hard to understand. It is helpful to ask your doctor about the cancer stage and how it will guide your treatment.

Order of treatments

Most people with soft tissue sarcoma will receive more than one type of treatment. You may want more information on when and why treatments are given. Talk to your doctor about the order of your treatment plan. The terms that describe the order of treatments are:



Neoadjuvant treatment is given before surgery to shrink the tumor.



Primary treatment is the main treatment given to rid the body of cancer.



Adjuvant treatment is given after surgery to kill any remaining cancer cells.

Stage I sarcoma

Guide 3 lists the treatment options for stage I sarcoma. Stage I is further grouped into stage IA and stage IB. Surgery to remove the tumor is recommended for primary treatment. For stage I, the intent of surgery is to cure the cancer.

Removing the sarcoma with a cancer-free surgical margin is the goal. However, your surgeon may remove a thin surgical margin to avoid cutting nerves and blood vessels. In this case, there is a higher chance that cancer cells will be left behind. A pathologist will assess the surgical margin for cancer cells. If cancer is found, you may have a second surgery.

Recommendations for more treatment are based on 1) the size of the margin; and 2) if the surgical cut went deep into a normal soft tissue layer called the fascia. Treatment options are based on whether the surgical margin is 1 cm or less, and if the fascia was cut.

Guide 3. Stage I sarcoma

The results of primary treatment are important. After surgery, your doctors will assess if you need further treatment. Based on the margins this could mean more surgery for a positive margin that does not include bone, major blood vessels, or nerves. Doctors may suggest observation after primary treatment. Observation is a period of scheduled follow-up testing to watch for signs of cancer spread (metastasis) or return (recurrence). Another option is radiation therapy to lower the chances of cancer returning.

Follow-up care will begin when treatment is complete. These tests will check for signs of cancer.

Stage		Primary treatment		Next treatment		Follow-up care
<ul style="list-style-type: none"> • Stage IA (T1, N0, M0, low grade) • Stage IB (T2–4, N0, M0, low grade) 	→	<ul style="list-style-type: none"> • Surgery, surgical margins >1 cm done or fascia not cut 		→		<ul style="list-style-type: none"> • See Guide 4
	→	<ul style="list-style-type: none"> • Surgery, surgical margins ≤1 cm and fascia cut 	→	<ul style="list-style-type: none"> • Surgery again • Observation (stage IA) • Consider radiation therapy 	→	<ul style="list-style-type: none"> • See Guide 4

Guide 4 lists follow-up care for after cancer treatment has ended. You should receive rehabilitation if you need it. This may include occupational therapy to help with daily life skills, or physical therapy to help your body move and function.

Follow-up tests are done at certain times to check if the cancer has returned. Getting follow-up tests can help find cancer early.

The follow-up tests include a medical history, physical exam, and imaging tests. For example, if the cancer is likely to spread to your lungs, you may get imaging tests of the chest. Likewise, you may get imaging tests of the site where the primary tumor was. Ask your doctor about what follow-up care you may need.

Guide 4. Follow-up care

Follow-up care

- Rehabilitation, if needed
- Medical history and physical exam every 3–6 months for 2–3 years, then repeat every year
- Consider imaging of chest every 6–12 months
- Consider imaging of primary tumor site right after surgery
- Consider regular imaging of primary tumor site

Stages II and III sarcoma

Treatment options for sarcoma are based on whether surgery can be done. Some sarcomas can't initially be removed by surgery because they are too large. Removing some sarcomas would limit use of your limb or other body part. Surgery may also not be an option because of your health. Treatment options for when surgery may be an option are listed next, followed by options for when surgery may not be possible.

Guide 5 has treatment options for stage II sarcomas that may be treated with surgery. For stage II, surgery alone may be an option if the tumor is small and the surgeon can remove the tumor with wide enough margins. The second option is surgery followed by radiation therapy. You should receive

radiation if cancer is found close to or at the margins and a second surgery can't be done.

The third option is to have radiation therapy followed by surgery. Radiation therapy before surgery may reduce the tumor size for surgery and lower the chances of the cancer returning. It may also improve how well your limb works after surgery. However, radiation will likely slow healing of the surgical wound. Before having surgery, you may get another imaging test. This test will assess the tumor and rule out metastatic disease.

Guide 6 shares options for stages IIIA and IIIB sarcoma. All options include surgery to remove the tumor. Multiple treatments can be combined with surgery. These other treatment options include radiation therapy, chemoradiation, or chemotherapy.

Guide 5. Stage II

Surgery may be an option

Stage	Treatment options
Stage II (T1, N0, M0, grade 2–3)	<ul style="list-style-type: none"> • Surgery • Surgery followed by radiation therapy • Radiation therapy followed by surgery

Guide 6. Stage III

Surgery may be an option

Stage	Treatment options
<ul style="list-style-type: none"> • Stage IIIA (T2, N0, M0 grade 2–3) • Stage IIIB (T3–4, N0, M0, grade 2–3) 	<ul style="list-style-type: none"> • Surgery followed by radiation therapy ± adjuvant chemotherapy • Chemoradiation or radiation therapy followed by surgery, then radiation therapy boost ± adjuvant chemotherapy • Chemotherapy followed by surgery, then radiation therapy ± adjuvant chemotherapy

All of these options may be given before surgery (preoperative) or after surgery (postoperative).

Neoadjuvant treatment may include chemoradiation, radiation therapy, or chemotherapy. The goal is to shrink the tumor before surgery. For example, NCCN experts prefer radiation therapy be given before surgery, especially if there is a large amount of disease.

After surgery for IIIA or IIIB disease, you may receive radiation therapy with or without adjuvant chemotherapy. A radiation boost may also follow surgery with prior chemoradiation or radiation therapy. A boost may help if the cancer wasn't fully removed or cancer was found in the margins.

A PET/CT may be used to find out if the cancer has responded to the chemotherapy. It is not exactly clear if adjuvant chemotherapy is helpful. It is still being studied in clinical trials. Your doctor may suggest joining a clinical trial that is testing adjuvant chemotherapy.

Guide 7 includes follow-up care after cancer treatment has ended. You should receive rehabilitation if needed. This may include occupational or physical therapy. You should also start to have follow-up tests to check if the cancer has returned.

Follow-up tests include a medical history, physical exam, and possible imaging of your chest. You may have imaging tests of the primary site right after surgery. If the cancer is likely to return, you may have regular imaging tests of the site where the primary tumor was. You may not have an imaging test if the area can be followed by physical exam.

Guide 7. Follow-up care

Follow-up care

- Rehabilitation, if needed
- Medical history, physical exam, and imaging of chest every 3–6 months for 2–3 years, then every 6 months for 2 years, then repeat every year
- Consider imaging of primary tumor site right after surgery
- Consider regular imaging of primary tumor site

Guide 8 lists the treatment options for stages II and III where surgery may not be an option. However, other treatments may shrink the tumor so that surgery can be done. These treatments include radiation therapy, chemoradiation, chemotherapy, or isolated limb infusion/perfusion.

Radiation treatment would be EBRT and the treatment planning should use IMRT, tomography, or protons. Isolated limb infusion/perfusion is another option to treat sarcoma of the limb. The drugs are injected into blood vessels within the limb. 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.

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. After surgery, you may have radiation therapy if you had none before. If you had radiation therapy before surgery, you may receive a radiation boost after surgery.

You may or may not have chemotherapy after surgery. It is not clear if adjuvant chemotherapy is helpful. A PET/CT may be used to find out if the cancer has responded to the chemotherapy. Joining a clinical trial that is testing chemotherapy may also be a good option.

Guide 8. Stages II and III sarcoma

Surgery may not be an option

Stage	Primary treatment	Treatment results	Next treatment
Stages II and III	<ul style="list-style-type: none"> • Radiation therapy • Chemoradiation • Chemotherapy • Regional limb therapy 	<p>→ Able to have surgery with good results</p>	<p>→</p> <ul style="list-style-type: none"> • Surgery + radiation therapy (if none before) ± adjuvant chemotherapy • Surgery + radiation therapy boost ± adjuvant chemotherapy
		<p>→ Unable to have surgery or good results</p>	<p>→</p> <ul style="list-style-type: none"> • Radiation therapy (if none before) • Chemotherapy • Palliative surgery • Observation if no symptoms • Best supportive care • Amputation

If you have no symptoms from the cancer, you may undergo observation. Observation is a period of regular testing for cancer growth so treatment can be started if needed. If the cancer is causing symptoms palliative surgery or other best supportive care may be recommended for relief from symptoms.

Amputation may be the best option for some patients. Amputation is the removal of an arm or leg by surgery. You may have an amputation if the limb will not function after surgery. A patient may also prefer this option and doctors may agree that amputation is necessary. Before deciding on treatment, your treatment team will first consider the advances in treatment regarding limb reconstruction. They will also consider multiple treatment options.

Ask your doctor:

- Questions about your treatment options
- For a list of possible side effects of each option
- Which option seems best for your situation

Guide 9 lists care after cancer treatment has ended. You should receive rehabilitation if needed. This may include occupational or physical therapy. You should also start to have follow-up tests to check if the cancer has returned.

Tests include medical history, physical exam, and imaging of the primary tumor site and/or the chest. You may have imaging tests of the primary site right after surgery. If the cancer is likely to return, you may have regular imaging tests of the site where the primary tumor was. You may not have an imaging test if the area can be followed by physical exam.

Guide 9. Follow-up care

Follow-up care

- Rehabilitation, if needed
- Medical history, physical exam, and imaging of chest every 3–6 months for 2–3 years, then every 6 months for 2 years, then repeat every year
- Consider baseline and regular imaging of primary tumor site

Stage IV sarcoma

There is little research that shows what are the best treatments for stage IV sarcomas. Thus, clinical trials are the preferred treatment option. If you are not able to join a clinical trial, the general recommendations presented next may help. The treatment options are divided into stage IV confined cancer (limited to one organ) and widespread cancer.

Guide 10 lists treatment options for stage IV sarcomas that have spread to one organ and aren't too large. In these cases, local treatment may be used to treat the primary tumor. Local treatment options are the same as those listed for stages II and III.

There are five other options to consider. One option is surgery to remove the metastases. This surgery is called a metastasectomy. You may be given chemotherapy, before or after surgery, with or without radiation therapy. The other four options are ablation, embolization, SBRT, and observation.

Follow-up care should start after treatment is complete. You may receive rehabilitation if needed. This may include occupational or physical therapy. Tests include medical history, physical exam, and imaging of the chest or metastatic sites on a recommended schedule (see Guide 10). If the cancer is likely to return, you may have regular imaging tests of the site where the primary tumor was.

Guide 10. Stage IV confined cancer

Treatment options	Follow-up care
<p>Treatment as listed for stages II and III in Guides 5 and 6, and consider these options:</p> <ul style="list-style-type: none"> • Surgery to remove metastases ± chemotherapy before or after surgery, ± radiation therapy • Ablation • Embolization • SBRT • Observation 	<ul style="list-style-type: none"> • Rehabilitation, if needed • Medical history and physical exam every 2–6 months for 2–3 years, then every 6 months for 2 years, then repeat every year • Imaging of chest and metastatic sites every 2–6 months for 2–3 years, then every 6 months for 2 years, then repeat every year • Consider baseline and regular imaging of primary tumor site

Guide 11. Stage IV widespread cancer

Palliative treatment options	
<ul style="list-style-type: none"> • Chemotherapy • Radiation therapy/SBRT • Surgery • Observation if no symptoms 	<ul style="list-style-type: none"> • Supportive care • Ablation • Embolization

Guide 11 lists the treatment options for widespread sarcoma. Widespread means the cancer is in many places in the body. If you have no symptoms, observation is an option. Observation is a period of regular testing for cancer growth so treatment can be started if needed.

If you do have symptoms, you can receive palliative care. Palliative care does not try to cure the cancer; it aims to relieve your discomfort. Chemotherapy, radiation therapy, and surgery may reduce cancer symptoms by stopping or slowing tumor growth. Other options including ablation and embolization may also be considered.

Recurrence treatment

Disease recurrence

It is possible for sarcoma to come back after cancer treatment is finished and the sarcoma is in remission. Remission is a period of being free of cancer. If sarcoma does come back, it may return in certain areas. It may be local (in or close to where it started), in one organ, in lymph nodes, or widespread.

Recurrent treatment options are shown in **Guide 12**. The treatment options are guided by where the cancer has returned. For example, a local recurrence may be treated like a newly diagnosed soft tissue sarcoma.

Guide 12. Recurrence treatment

Cancer spread	Treatment options
Local recurrence	<ul style="list-style-type: none"> • Testing, then treatment based on stage (extent of disease) See Guides 2–8
Tumor in one organ and it's not large	<ul style="list-style-type: none"> • Surgery to remove metastases ± chemotherapy before or after surgery ± radiation therapy • Ablation • Embolization • SBRT
Widespread recurrence	<ul style="list-style-type: none"> • Palliative chemotherapy • Palliative radiation therapy/SBRT • Palliative surgery • Observation if no symptoms • Supportive care • Palliative ablation • Palliative embolization
Isolated regional disease or lymph nodes	<ul style="list-style-type: none"> • Regional node dissection ± RT ± chemotherapy • Surgery to remove metastases ± chemotherapy before or after surgery ± radiation therapy • SBRT • Isolated limb infusion/perfusion + surgery

Guide 12 lists the treatment options for a recurrence of sarcoma. A recurrence means the cancer has come back after being disease-free for a period of time. Options are listed by the extent of the recurrence. The first list of treatment options is for a local recurrence of the primary tumor. Options are based on the stage or extent of the local recurrence. Testing and treatment are listed in Guides 2 through 8, earlier in this chapter.

Secondly, sarcoma may return as a small tumor within one organ. In this case, surgery to remove the tumor is an option. You may receive chemotherapy before or after surgery, with or without radiation therapy. Other options include ablation, embolization, or SBRT.

The third list of treatment options is for widespread sarcoma. Treatment options depend on whether or not the cancer is causing symptoms. If you have no symptoms, observation is an option.

If you do have symptoms, you can receive palliative care. Palliative care is also called supportive care. Palliative care does not try to cure the cancer. It aims to relieve your discomfort. Chemotherapy, radiation therapy, and surgery may reduce cancer symptoms by stopping or reducing tumor growth. Ablation, embolization, and SBRT may also reduce symptoms.

The last set of options is for isolated regional disease or lymph nodes. The cancer has returned near the site of the primary tumor or lymph nodes close to where the cancer started. The removal of lymph nodes is an option. You may or may not have radiation therapy with or without chemotherapy.

Another option is surgery to remove the metastases. You may be given chemotherapy, before or after surgery, with or without radiation therapy. The other two options are SBRT, or an isolated limb infusion/perfusion and surgery.

For an isolated limb infusion/perfusion, chemotherapy is infused into the arm or leg during a surgical procedure. This procedure should be done at hospitals or centers with experience using this treatment.

Review

- Treatment planning is an important first step of care.
- Most stage I sarcomas in the limbs, outer trunk, head, or neck can be treated with surgery.
- Stages II and III sarcomas may or may not be treated with surgery.
- Treatment for stage IV sarcomas depends on whether the metastases are in one organ or widespread.
- Treatment options for recurrences are based on the extent of the cancer.

5

Treatment guide

Sarcomas in the inner trunk

-
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Part 5 is about sarcomas within the retroperitoneal, abdominal, or pelvic spaces. We refer to this as the inner trunk. This chapter starts by listing what is needed for treatment planning. Treatment is based on whether surgery is an option for the sarcoma in the inner trunk.

This information is taken from the treatment guidelines written by NCCN experts of sarcoma. These treatment guidelines list options for people with sarcoma in general. Thus, your doctors may suggest other treatment for you based on your health and personal wishes.

Treatment planning

Guide 13 shares what tests are needed prior to starting treatment. A team of experts who have experience with sarcoma should assess the extent of disease and make a treatment plan. This plan will focus on sarcoma in the inner trunk. In order to make a plan, certain tests will be done to find out more about your health and medical needs. Your doctors will take your medical history, do an exam of your body, order imaging tests, and may do a biopsy. A biopsy is the clear way to confirm a sarcoma.

If you have a personal or family history of a rare hereditary cancer syndrome called Li-Fraumeni syndrome (see page 15), NCCN experts suggest a genetics assessment. This assessment includes meeting with a genetic counselor who can talk with you about inherited diseases such as Li-Fraumeni syndrome.

Guide 13. Tests for sarcoma in the inner trunk

Needed for most patients	May be needed for some patients
<ul style="list-style-type: none"> • Medical history and physical exam 	<ul style="list-style-type: none"> • Biopsy before surgery
<ul style="list-style-type: none"> • CT of chest, abdomen, and pelvis ± MRI of abdomen and pelvis 	<ul style="list-style-type: none"> • Genetic assessment
<ul style="list-style-type: none"> • Biopsy if: <ul style="list-style-type: none"> ◦ You will have neoadjuvant treatment ◦ Your doctor wants to assess for other cancer besides a sarcoma 	

Treatment with surgery

Guide 14 lists the treatment options for sarcomas that can be treated with surgery. Surgery with or without IORT is an option whether or not you had a biopsy. If you had a biopsy, and you do not have a GIST or desmoid tumor, radiation therapy or chemotherapy before surgery is another option. The goal is to remove all the cancer with cancer-free surgical margins.

Treatment options will depend on the type of sarcoma found during the biopsy or surgery. Once you have the results, it is helpful to follow the arrows in Guide 14. Refer to Guide 15 on the next page for adjuvant treatment options. Adjuvant treatment comes after a primary treatment like surgery.

Guide 15 (on the next page) shows the possible results of surgery and next steps of care. After surgery, your doctors will assess if you need further treatment such as adjuvant treatment. Adjuvant treatment may kill any remaining cancer cells and help stop the cancer from returning.

Recommendations for more treatment are based on whether cancer is at the surgical margin. A pathologist will assess the surgical margin for cancer cells. Many people will not have more treatment if no cancer remains. When no disease remains, only some patients will get radiation therapy. This is because the cancer is likely to return. Others with no disease will move on to follow-up care with testing on a regular schedule.

Guide 14. Treatment with surgery

Biopsy		Results		Treatment options
Biopsy	→	• GIST and desmoid tumors	→	• See Part 5 for GIST, and Part 6 for desmoid tumors
		• Other sarcoma	→	• Surgery ± IORT • Chemotherapy or radiation before surgery, then surgery ± IORT
No biopsy, surgery	→	• GIST and desmoid tumors	→	• See Part 5 for GIST, and Part 6 for desmoid tumors
		• Other sarcoma	→	• See Guide 15

Some margins have cancer cells that aren't seen with the naked eye. Instead, cancer cells are found with a microscope. Thus, some disease remains after surgery. In this case, you may have radiation therapy to kill any remaining cells. However, if you had radiation therapy before surgery, only a radiation boost may be given.

Often, cancer can still be seen in the surgical margin after the sarcoma is removed. This occurs because the tumor is close to important structures. Examples of such structures include large blood vessels or nerves. A second surgery to remove the remaining cancer may be an option. If surgery isn't possible, there are other treatments. See Guide 16 on the next page for more information.

Guide 15 also shares follow-up tests after cancer treatment has ended. You should have follow-up tests to check if the cancer has returned. Follow-up tests include a physical exam and imaging of your abdomen and pelvis. If the cancer is likely to spread to your lungs, you may get imaging tests of the chest. Certain tests follow a schedule while others are based on any signs or symptoms you have during follow-up care.

Guide 15. Treatment after surgery and follow-up care

Results of surgery		Adjuvant treatment		Follow-up tests
No disease remains	→	<ul style="list-style-type: none"> Consider radiation therapy only for certain patients 	→	<ul style="list-style-type: none"> Physical exam Imaging of abdomen and pelvis <ul style="list-style-type: none"> Every 3–6 months for 2–3 years then every 6 months for 2 years, then repeat every year Consider imaging of chest
Some disease remains	→	<ul style="list-style-type: none"> Consider radiation therapy only for certain patients Consider radiation boost for certain patients if received radiation before 	→	
Disease remains	→	<ul style="list-style-type: none"> Consider surgery if possible Consider other treatments if surgery not possible 	→	

Treatment without surgery

Guide 16 lists the treatment options for stage IV disease or sarcomas that may not have surgery as the first option. For some people, other treatments may shrink the tumor so that surgery may be done later on. These other treatments include radiation therapy, chemotherapy, or both combined. This is called chemoradiation. Radiation may be given if the cancer was never treated with it before. If after these treatments you are able to have surgery, see Guide 14 on page 43. If you are unable to have surgery, palliative care is an option to control the symptoms.

Chemotherapy, radiation therapy, and surgery may stop or decrease tumor growth. Palliative care options depend on your symptoms. If you have no symptoms, observation is an option. Your doctor may also suggest surgery to remove metastatic tumors that are causing symptoms. Joining a clinical trial is also an option and is highly encouraged. Ask your treatment team if there are clinical trials you can join.

Review

- Treatment planning is an important first step of care.
- Sarcomas in the inner trunk may or may not be treated with surgery.
- Follow-up tests include a physical exam and imaging of your chest, abdomen, and pelvis.

Guide 16. Treatment for stage IV or without surgery as primary treatment Surgery may not be the first treatment option

Treatment options	Treatment response		Next steps
Consider to down-stage disease: <ul style="list-style-type: none"> • Chemotherapy • Chemoradiation • Radiation therapy 	➔	<ul style="list-style-type: none"> • Imaging to assess response to treatment 	➔
			➔
Palliative care only	➔		<ul style="list-style-type: none"> • Surgery not an option or disease progressing: <ul style="list-style-type: none"> ◦ Chemotherapy ◦ Radiation therapy ◦ Surgery for symptom control ◦ Supportive care ◦ Observation if no symptoms

6

Treatment guide

Gastrointestinal stromal tumors

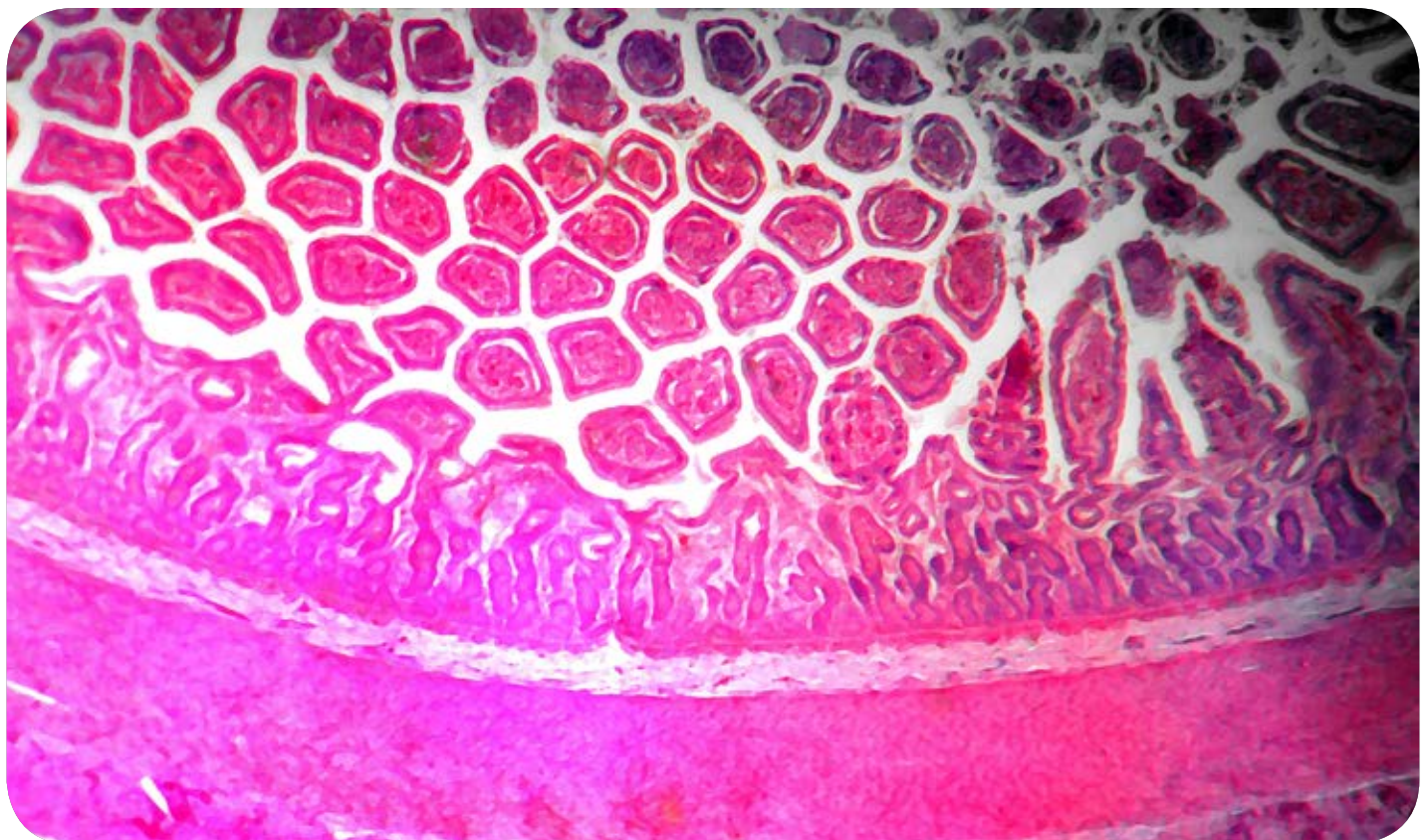
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Part 6 is about GISTs (gastrointestinal stromal tumors). These tumors may start in cells that trigger muscle movement in the gut wall. This chapter starts with treatment planning. Next, treatment options are presented for small stomach GISTs, other GISTs, and GISTs that keep growing while taking drug treatments.

This information is taken from the treatment guidelines written by NCCN experts of sarcoma. These treatment guidelines list options for people with sarcoma in general. Thus, your doctors may suggest other treatment for you based on your health and personal wishes.

Treatment planning

Guide 17 lists tests used to diagnose (confirm) GIST. A team of experts who have experience with sarcoma should assess the extent of disease and create a treatment plan for GIST. Your doctors will take a medical history, do an exam of your body, order imaging tests, and do a biopsy.

For a small GIST tumor of the stomach less than 2 cm, imaging should include the abdomen and pelvis, but not the chest. The biopsy for these tumors will be an EUS-FNA (endoscopic ultrasound-guided fine-needle aspiration). This type of biopsy is preferred. It removes a sample of the tumor with a needle on an imaging device guided through a natural opening of the body.

Your doctor may also suggest tests for gene mutations. Genes are the instructions in cells for making new cells and controlling how cells behave. An abnormal change in these instructions—called a gene mutation—can cause cells to grow and divide out of control. Gene mutations in GIST may include *KIT*, *PDGFRA* (platelet-derived growth factor receptor alpha), and *SDH*.

Genotyping may be a part of treatment planning. Genotyping looks at your genetic make-up. This is useful because some drugs target certain genes associated with the cancer. Mutations in *KIT* and *PDGRFA* can affect how the sarcoma reacts to targeted therapy. People with an *SDH* mutation may not be able to take imatinib, but there are other treatment options. Thus, the tests in Guide 17 are important.

Guide 17. Tests for GIST

Needed for most patients	May be needed for some patients
<ul style="list-style-type: none"> Medical history and physical exam 	<ul style="list-style-type: none"> Imaging of chest
<ul style="list-style-type: none"> Biopsy 	<ul style="list-style-type: none"> EUS-FNA
<ul style="list-style-type: none"> CT ± MRI of abdomen and pelvis 	<ul style="list-style-type: none"> Testing for gene mutations
	<ul style="list-style-type: none"> Genotyping for treatment planning

The results will help your treatment team learn more about the GIST tumor and make a treatment plan.

for 3 to 5 years. If test results are normal for 3 to 5 years, you will have a CT scan every year.

Treatment for small stomach GISTs

Guide 18 shows the treatment options for GISTs of the stomach that are less than 2 cm. Many of these tumors are benign (not cancer). They are usually not aggressive. However, some will grow fast and may become a problem.

Tumors that aren't high risk don't need treatment. Instead, the tumor can be followed for growth by endoscopic or imaging tests.

Your doctors will assess the tumor. They will look for risk factors that suggest there is a high risk for fast growth. Surgery to remove the tumor is recommended for those who are at high risk of the tumor growing fast.

After surgery, your doctor will follow you closely. You may get CT scans of your abdomen and pelvis. It is suggested the CT scans occur every 3 to 6 months

What to do...

- ✓ Ask questions about your test results.
- ✓ Learn more about the treatment options your team recommends.
- ✓ Find out what side effects are possible during and after treatment.

Guide 18. Treatment for small stomach GISTs

Less than 2 cm

EUS-FNA test results		Primary treatment		Tests
High-risk features	→	• Surgery	→	<ul style="list-style-type: none"> • Medical history and physical exam • Consider CT of abdomen and pelvis every 3–6 months for 3–5 years, then repeat every year
No high-risk features		→		<ul style="list-style-type: none"> • Consider regular endoscopic or imaging tests

See Part 2 on page 11 to learn more about these tests. Ask your doctors about the benefits and possible risks of these tests.

Treatment for other GISTs

Guide 19 lists the treatment options for all GISTs other than small stomach GISTs. Options are listed by whether or not a tumor can first be treated with surgery. Tumors that can first be treated with surgery have not grown or spread far. Larger tumors, cancer that has returned after surgery, and metastatic disease are often first treated with imatinib.

Surgery first

Surgery can be your first treatment if it does not put you at unreasonable risk for complications. Your surgeon may remove nearby organs along with the tumor. Some GIST tumors (in the front wall of stomach and parts of the small intestine) may be removed by laparoscopic surgery. This method uses small cuts to do work inside the belly area.

After any type of surgery, you may receive more treatment. If all the cancer appears to be removed, you could start imatinib if there's a high or medium risk for the cancer returning. If your surgeon was unable to remove all of the cancer, you may start imatinib and consider a second surgery.

Guide 19. Treatment for other GISTs

Possible surgery		Treatment options		Treatment results		Next treatment
Surgery is an option	→	<ul style="list-style-type: none"> • Surgery • Imatinib, then surgery (if high risk of issues after surgery) 		• All cancer removed	→	<ul style="list-style-type: none"> • Imatinib: <ul style="list-style-type: none"> ◦ If medium/high risk of recurrence (and had none before) ◦ If taken before surgery • Observe if low risk of recurrence
				• Not all cancer removed	→	• Imatinib and consider second surgery
				• Metastases	→	• Imatinib
Surgery is not the best first option	→	• Imatinib	→	• Response or stable disease	→	• Continue imatinib and consider surgery
				• Progression	→	• See Guide 21

Imatinib first

Imatinib is often used first for GISTs that can be removed by surgery but the surgery would leave you with long-term problems or the surgery could be made simpler by shrinking the tumor. Imatinib is also used for GISTs that can't be removed by surgery, returned after surgery, or have spread far. It is very important that you don't stop taking imatinib once you've started.

After taking imatinib for a few weeks, a PET scan or a CT scan can show if treatment is working. If the tumor is less active on PET scan or the same size or smaller on CT scan, keep taking the same dose of imatinib. Once the tumor is small enough, you may be able to have surgery. Your surgeon and medical oncologist should assess if surgery is possible.

After surgery, start imatinib again for adjuvant treatment. If the tumor grows while taking imatinib, surgery may be done for some people. Otherwise, see Guide 21, on the next page, for treatment options.

Guide 20 lists follow-up care for after surgery. You should have follow-up tests. Test recommendations are based on surgery results. The surgery may have removed all the cancer or some cancer may remain. If all the cancer was removed, testing can assess if the cancer has returned. Getting follow-up tests can help find cancer early. To find cancer early, get a medical history and physical exam every 3 to 6 months for 5 years. If results are normal for 5 years, then these tests may be done every year. In addition, get a CT of your abdomen and pelvis. If results are normal during this time, the follow-up tests are done once a year.

Guide 20. Follow-up care after surgery

Surgery results	Follow-up care
All cancer removed	<ul style="list-style-type: none"> • Medical history and physical exam every 3–6 months for 5 years, then repeat every year • Imaging of abdomen and pelvis every 3–6 months for 3–5 years, then repeat every year
Not all cancer removed (persistent or metastatic disease)	<ul style="list-style-type: none"> • Medical history, physical exam, and imaging of abdomen and pelvis every 3–6 months

If not all the cancer was removed, testing can assess if the cancer is growing. Tests should include medical history, physical exam, and imaging tests of your abdomen and pelvis every 3 to 6 months. If the cancer is growing, read the next section, *Treatment for disease progression*.

Treatment for disease progression

Some GISTs grow while taking drug treatment. Doctors call this cancer progression. Treatment options after progression are presented next.



Guide 21 lists treatment options based on whether there is limited or widespread growth. If limited growth, you may keep taking imatinib and consider

local treatments. Otherwise, you may increase the dose of imatinib or start taking sunitinib. Sunitinib may be given if the tumor does not respond to imatinib or the side effects of imatinib are too intense. Thus, you will need to stop taking imatinib.

For widespread cancer growth, options are to increase the dose of imatinib or start taking sunitinib. After the new treatment, your doctors will assess if the treatment is working with CT or MRI. PET/CT may be used if CT or MRI results are unclear.

Joining a clinical trial is highly encouraged. You will likely have the best management of care. Ask your treatment team if there are clinical trials you can join. See page 27 to learn more about clinical trials.

Guide 21. Treatment for disease progression

Cancer growth		Treatment options
Limited growth		<ul style="list-style-type: none"> • Continue imatinib and consider: <ul style="list-style-type: none"> ◦ Surgery if possible ◦ Ablation, embolization, or chemoembolization ◦ Palliative radiation therapy if cancer spread to the bones • Increase dose of imatinib or change to sunitinib and do imaging tests to assess treatment response
Widespread growth		<ul style="list-style-type: none"> • For certain patients: <ul style="list-style-type: none"> ◦ Increase dose of imatinib or change to sunitinib and do imaging tests to assess treatment response

Guide 22 lists treatment options if the cancer keeps growing while taking imatinib or sunitinib. There are four treatment options. Taking regorafenib is the first option. You may also join a clinical trial or start another treatment. However, less research is able to support these other treatments at this time. The fourth option is to receive supportive care to relieve any discomfort you may have. As part of supportive care, you should continue to take imatinib or another drug even if your tumor is growing. This may slow the tumor growth.

Review

- GISTs may start in cells that trigger muscle movement in the gut wall.
- Small stomach GISTs that likely won't grow fast don't need treatment. Instead, these tumors may be watched for cancer growth.
- Small stomach GISTs that likely will grow fast can be removed with surgery.
- All other GISTs first may be treated with surgery or imatinib.
- If GISTs grow while you're taking imatinib, the dose may be increased or you may be switched to sunitinib.

Guide 22. Treatment for progression that continues

Treatment options

For disease that progresses despite treatment with imatinib or sunitinib, consider these options:

- Regorafenib
- Clinical trial
- Consider these other options:
 - Sorafenib
 - Nilotinib
 - Dasatinib (for patients with *D842V* mutation)
 - Pazopanib
 - Everolimus + TKI (tyrosine kinase inhibitor)
- Best supportive care

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Treatment guide

Desmoid tumors

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Rhabdomyosarcoma

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Part 7 starts with treatment planning for desmoid tumors. Treatment options are then presented for when surgery can and can't be done for desmoid tumors. The next section focuses on treatment options for rhabdomyosarcoma. These treatment options include systemic therapies.

This information is taken from the treatment guidelines written by NCCN experts of sarcoma. These treatment guidelines list options for people with sarcoma in general. Thus, your doctors may suggest other treatment options for you based on your health and personal wishes.

Desmoid tumors

Desmoid tumors tend to be benign (not cancer), although they can act like cancer. Desmoid tumors are also known as aggressive fibromatoses. Thus, some doctors refer to them as “low-grade sarcoma.” They can grow into nearby tissue but won't spread to distant sites. There is also a high risk for local recurrence of these tumors. Many factors, including the type of tumor, whether there is other sarcoma

found, and the location are all a part of planning treatment. Surgery is the main treatment for desmoid tumors that can be removed.

Treatment planning

Guide 23 shows some tests used for desmoid tumors. A team of experts who have experience with sarcoma should assess the extent of disease. In order to make a plan, certain tests will be done to find out more about your health. Your doctors will take a medical history, do an exam of your body, order imaging tests, and do a biopsy. A biopsy may not be needed if surgery is planned.

Desmoid tumors often occur for unknown reasons. However, some people have syndromes that increase their chances of getting these tumors. Gardner's syndrome and FAP are such syndromes. Both are rare disorders that increase the chances for cancer and desmoid tumors. Your doctors will assess if you likely have either of these syndromes. A screening colonoscopy may be recommended for some people.

Guide 23. Tests for desmoid tumors

Needed for most patients	May be needed for some patients
<ul style="list-style-type: none"> Medical history and physical exam <ul style="list-style-type: none"> Assess for Gardner's syndrome or FAP 	<ul style="list-style-type: none"> Biopsy
<ul style="list-style-type: none"> Imaging of tumor 	

Treatment with surgery

Guide 24 lists the treatment options for desmoid tumors that can be treated with surgery. Some tumors are small enough, aren't causing symptoms, and are located where an increase in size won't cause problems. For these tumors, you may start observation. Observation is a period of testing to watch for tumor growth so that treatment can be started if needed.

Large tumors that are causing problems should be treated based on where the tumor is located and the possible outcomes of treatment. Options include surgery or radiation therapy with or without drug treatment using systemic therapy.

You may receive more treatment after primary treatment. If the tumor is all gone, you may start observation. For large tumors, radiation therapy after surgery can be given if you did not receive it before surgery. There are three options if most of the tumor was removed but tiny amounts that can't be seen remain. These options are observation or a second surgery.

Tumors that can still be seen after treatment have four options for treatment. Radiation and systemic therapy are two options. If neither option works, surgery to remove the visible tumor may be done. Observation may also be an option. If the tumor grows, you can start treatment.

Guide 24. Treatment with surgery

Primary treatment		Treatment results		Next treatment or other care
Observation for certain patients	→	• Stable	→	<ul style="list-style-type: none"> • Continue observation • Medical history and physical exam with imaging tests
		• Progression	→	<ul style="list-style-type: none"> • Follow treatment paths below
Surgery	→	• No disease remains	→	<ul style="list-style-type: none"> • Observation
		• Some disease remains and can't be seen	→	<ul style="list-style-type: none"> • Observation • Consider another surgery
		• Disease remains and can be seen	→	<ul style="list-style-type: none"> • Radiation therapy • Systemic therapy • Consider surgery if other treatment doesn't work • Observation
Radiation therapy and/or systemic therapy			→	<ul style="list-style-type: none"> • See Guide 25

Systemic therapies for desmoid tumors may include sulindac or other NSAIDs including celecoxib, tamoxifen with or without sulindac, toremifene, methotrexate and vinblastine, low-dose interferon, doxorubicin-based regimens, imatinib, sorafenib, methotrexate and vinorelbine, and liposomal doxorubicin. Ask your doctor if systemic therapies are an option for you. See Guide 1 on pages 24 and 25.

Guide 25 refers to follow-up care after surgery or radiation therapy and/or systemic therapy has ended. You should receive rehabilitation if needed. This may include occupational or physical therapy. Report any new or worse symptoms to your doctors. A change in symptoms may be a sign of tumor growth. You

should also start to have follow-up tests to check if the tumor has returned. Tests include medical history, physical exam, and imaging. These tests should be done every 3 to 6 months for 2 to 3 years, then every 6 to 12 months.

Treatment without surgery

Guide 26 lists recommended options for tumors that can't be fully removed with surgery. There are four treatment options. Radiation and systemic therapy are two options. If neither option works, surgery to remove the visible tumor may be done.

Guide 25. Follow-up care and treatment for progression or recurrence

Follow-up care	Treatment options for progression or recurrence
<ul style="list-style-type: none"> • Monitor for symptoms • Rehabilitation, if needed • Medical history, physical exam, and imaging every 3–6 months for 2–3 years, then every 6–12 months 	<ul style="list-style-type: none"> • Systemic therapy • Surgery • Surgery + radiation therapy • Radiation therapy (if none before)

Guide 26. Treatment without surgery and follow-up care

Primary treatment	Follow-up care	Progression or recurrence
<ul style="list-style-type: none"> • Radiation therapy • Systemic therapy • Consider surgery if other treatment doesn't work • Observation 	<ul style="list-style-type: none"> • Rehabilitation, if needed • Medical history, physical exam, and imaging every 3–6 months for 2–3 years, then every 6–12 months 	<ul style="list-style-type: none"> • See primary treatment options

Observation may also be an option. If the tumor grows, you can start treatment.

Follow-up care will start after treatment is complete. You should receive rehabilitation if needed. This may include occupational or physical therapy. Report any new or worse symptoms to your doctors. A change in symptoms may be a sign of tumor growth.

You should also start to have follow-up tests to check if the tumor has returned. Tests include medical history, physical exam, and imaging. These tests should be done every 3 to 6 months for 2 to 3 years. If results are normal during this time, these tests may be done every 6 to 12 months.

Review

- ▶ Desmoid tumors are also known as aggressive fibromatoses.
- ▶ Some desmoid tumors can be treated with surgery while others receive other treatment.
- ▶ Receiving care after treatment is important to find any growing or new tumors early.

Rhabdomyosarcoma

RMS (rhabdomyosarcoma) is a rare type of sarcoma that usually affects children. RMS has three subtypes: embryonal (including botryoid and spindle cell variants), alveolar (including a solid variant), and pleomorphic histologies.

For this section, treatment guides are separated into pleomorphic RMS, which mostly occurs in adults, and non-pleomorphic RMS. Since this cancer is rare in adults, the data is mostly based on treating children with RMS. Pleomorphic RMS mostly occurs in the limbs, trunk wall, urinary system (kidneys, bladder, urethra, and ureter), and reproductive (vagina, uterus, testes, etc.) system. Treatment may involve surgery, radiation therapy, and chemotherapy.

Treatment for RMS

[Guide 27](#) on page 58 has treatment options for RMS. PET or PET/CT scan may be helpful in assessing the extent of disease. Your doctor can check the lymph nodes and other sites for metastatic disease. Treatment planning should include a group of experts who treat RMS. More than one treatment may be offered. These include surgery, radiation therapy, and chemotherapy.

The pleomorphic type of RMS is more common in adults and may not have the same treatment options as other soft tissue sarcomas. Treatment options include systemic therapy as seen in [Guide 28](#) on page 58. This list has combinations of drugs and single agents recommended by NCCN experts, with more than one treatment being considered for adults with RMS.

Guide 27. Treatment for RMS

Type of RMS		Treatment and other care
Pleomorphic RMS	→	<ul style="list-style-type: none"> • Treat like soft tissue sarcoma, see Guides 3–12 <ul style="list-style-type: none"> ◦ Consider systemic therapy, see Guide 28
Non-pleomorphic RMS (includes alveolar and embryonal)	→	<ul style="list-style-type: none"> • Seek expert care from a specialist who treats RMS • Pediatric, medical, surgical, and radiation oncologists should do evaluation • Assess risk and use multiple treatment options including systemic therapy in Guide 29

Guide 28. Systemic therapy for pleomorphic RMS

Combination regimens	Single agent
• AD (doxorubicin, dacarbazine)	• Doxorubicin
• AIM (doxorubicin, ifosfamide, mesna)	• Ifosfamide
• MAID (mesna, doxorubicin, ifosfamide, dacarbazine)	• Epirubicin
• Ifosfamide, epirubicin, mesna	• Gemcitabine
• Gemcitabine and docetaxel	• Dacarbazine
• Gemcitabine and vinorelbine	• Liposome doxorubicin
• Gemcitabine and dacarbazine	• Temozolomide
	• Vinorelbine
	• Eribulin
	• Trabectedin
	• Pazopanib

For non-pleomorphic RMS, including alveolar and embryonal types, treatment should be planned by a team of specialists. A pediatric oncologist may be involved in your care since this type is mostly seen in children. Medical, radiation, and surgical oncologists may also weigh in on your treatment options. Your doctors can assess the risk of each option and help you decide on your next steps of care. This may include systemic therapy. There are limited data on chemotherapy for adult RMS, since most clinical trials have focused on children with RMS. NCCN experts have created a list of drug treatment options found in [Guide 29](#). These drugs have shown action in the treatment of non-pleomorphic RMS.

Review

- RMS is a rare type of sarcoma in adults. It is more common in children.
- Treatment planning for rhabdomyosarcoma should include a team of expert doctors who treat cancer. This team may include a pediatric (child) oncologist.
- People with RMS often require more than one type of cancer treatment including surgery, radiation therapy, and/or chemotherapy.

Guide 29. Systemic therapy for non-pleomorphic RMS

Combination regimens	Single agent
• Vincristine, dactinomycin, cyclophosphamide	• Doxorubicin
• Vincristine, doxorubicin, cyclophosphamide	• Irinotecan
• Vincristine, doxorubicin, cyclophosphamide (alternate with ifosfamide and etoposide)	• Topotecan
• Vincristine, doxorubicin, ifosfamide	• Vinorelbine
• Cyclophosphamide and topotecan	• High-dose methotrexate
• Ifosfamide and doxorubicin	• Trabectedin
• Ifosfamide and etoposide	
• Irinotecan and vincristine	
• Vincristine and dactinomycin	
• Carboplatin and etoposide	
• Vinorelbine and low-dose cyclophosphamide	
• Vincristine, irinotecan, temozolomide	

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Making treatment decisions

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Having cancer can feel very stressful. While absorbing the fact that you have cancer, you must also learn about tests and treatments. And, the time you have to decide on a treatment plan may feel short.

Parts 1 through 7 described sarcomas along with the tests and treatment options recommended by NCCN experts. Part 8 aims to help you make decisions and talk with your treatment team about your next steps of care. You will also find a list of websites where you can learn more, seek support, or get resources on sarcoma.

It's your choice

The role patients want in choosing their treatment differs. You may feel uneasy about making treatment decisions. This may be due to a high level of stress. It may be hard to hear or know what others are saying. Stress, pain, and drugs can limit your ability to make good decisions. You may feel uneasy because you don't know much about cancer. You've never heard the words used to describe cancer, tests, or treatments. Likewise, you may think that your judgment isn't any better than your doctors'.

Letting others decide which option is best may make you feel more at ease. However, whom do you want to make the decisions? You may rely on your doctors alone to make the right decisions. However, your doctors may not tell you which to choose if you have multiple good options. You can also have loved ones help. They can gather information, speak on your behalf, and share in decision-making with your doctors. Even if others decide which treatment you will receive, your treatment team may still ask that you sign a consent form.

On the other hand, you may want to take the lead or share in decision-making. In shared decision-making, you and your doctors share information, discuss the options, and agree on a treatment plan. Your doctors know the science behind your plan but you know your concerns and goals. By working together, you can decide on a plan that works for you when it comes to your personal and health needs.

Questions to ask

You will likely meet with experts from different fields of medicine. It is helpful to talk with each person. Prepare questions before your visit and ask questions if the information isn't clear. You can get copies of your medical records. It may be helpful to have a family member or friend with you at these visits to listen carefully and even take notes. A patient advocate or navigator might also be able to come. They can help you ask questions and remember what was said.

The questions in this chapter are suggestions for information you read about in this book. Feel free to use these questions or come up with your own personal questions to ask your doctor and other members of your treatment team.

Questions about testing and the results

1. What tests will I have for this type of cancer?
2. Where and when will the tests take place?
3. How long will they take?
4. How do I prepare for testing?
5. How soon will I know the results and who will explain them to me?
6. Where did the sarcoma start? What is the cancer stage?
7. What can you tell me about this type of sarcoma?
8. Have any cancer cells spread to other parts of my body?

Questions about treatment options

1. What treatment options do I have?
2. Can I join a clinical trial?
3. Will my treatment team include specialists who have experience with sarcoma?
4. Does this hospital or center offer the best treatment for me?
5. Can you provide me with the research that supports this treatment plan?
6. Will I need more than one treatment?
7. How much time do I have to think about my options?
8. Do I have time to get a 2nd opinion?
9. How do I prepare for treatment?
10. Will I have to go to the hospital or elsewhere? How often will I go?
11. Should I bring someone with me when I get treated?

Questions about clinical trials

1. What clinical trial is right for me?
2. How many people will be in the clinical trial?
3. What are the tests and treatments for this study? And how often will they be?
4. How long will I be in the clinical trial?
5. Will I be able to get other treatment if this doesn't work?
6. How will you know if the treatment is working?
7. Who will help me understand the costs of the clinical trial?

Questions about side effects

1. What are the side effects?
2. When can they start?
3. How long will the side effects last?
4. When should I call the doctor about my side effects?
5. How can I prevent or relieve these side effects?
6. Are there any complementary therapies that might help?
7. Are there any long-term side effects from this treatment?
8. Is home care after treatment needed? If yes, what type?

Questions about a doctor's experience

1. Are you board certified? If yes, in what area?
2. How many people like me have you treated?
3. How many procedures like the one you're suggesting have you done?
4. Is this treatment a major part of your practice?
5. How many of your patients have had complications?

Deciding between options

Deciding which option is best can be hard. Doctors from different fields of medicine may have different opinions on which option is best for you. This can be very confusing. Your spouse or partner may disagree with which option you want. This can be stressful. In some cases, one option hasn't been shown to work better than another, so science isn't helpful. Some ways to decide on treatment are discussed next.

Getting a 2nd opinion

Even if you like and trust your doctor, it is helpful to get a 2nd opinion. You will want to have another doctor review your test results. He or she can suggest a treatment plan or check the one you already heard about.

Things you can do to prepare:

- Check with your insurance company about its rules on 2nd opinions. You will want to know about out-of-pocket costs for doctors who are not part of your insurance plan.
- Reach out to patient advocacy organizations (see websites on page 68) for help with 2nd opinions. Some may give referrals to hospitals or cancer centers who specialize in treating sarcoma.
- Make plans to have copies of all your records sent to the doctor you will see for your 2nd opinion. Do this well before your appointment. If you run into trouble having records sent, pick them up and bring them with you.

If your new doctor offers different advice, make an appointment with your first doctor to talk about the differences. Do whatever you need to feel confident about your diagnosis and treatment plan.

Keep in mind...

- ✓ Every treatment option has benefits and risks. Consider these when deciding which option is best for you.
- ✓ Talking to others may help identify benefits and risks you hadn't thought of.

Getting support

Support groups often include people at different stages of treatment. They can be online or meet in person. Some people in a support group may be in the process of deciding while others may be finished with treatment.

At support groups, you can ask questions and hear about the experiences of other people with soft tissue sarcoma. If your hospital or community doesn't have support groups for people with soft tissue sarcoma, check out the websites on the next page. These organizations may give referrals to support groups in your area, have online support groups, or have free hotlines you can call for help.

You can also reach out to a social worker or psychologist. They can help you find ways to cope or refer you to support services. These services may also be available to your family, friends, and those with children so they can connect and get support.

Websites

American Cancer Society

cancer.org/cancer/soft-tissue-sarcoma.html

GIST Support International

gistsupport.org

National Cancer Institute

cancer.gov/cancertopics/pdq/treatment/adult-soft-tissue-sarcoma/Patient

Sarcoma Alliance

sarcomaalliance.org

Sarcoma Alliance for Research through Collaboration

sarctrials.org

Sarcoma Foundation of America

curesarcoma.org

Review

- ▶ Shared decision-making is a process in which you and your doctors plan treatment together.
- ▶ Asking your treatment team questions is vital to getting the information you need to make informed decisions.
- ▶ Getting a 2nd opinion, attending support groups, and comparing benefits and risks may help you decide which treatment is best for you.

Glossary

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Dictionary

abdomen

The belly area between the chest and pelvis.

ablation

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

adjuvant therapy

Treatment that is given to lower the chances of the cancer returning.

allergic reaction

An abnormal response by the body to a foreign substance.

angiogram

A test that uses x-rays to make pictures of blood flow within an artery.

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.

cancer grade

A rating of how much cancer cells look like normal cells.

cancer stage

A rating of the outlook of a cancer based on its growth and spread.

Carney-Stratakis syndrome

A rare health condition that increases the chance of gastrointestinal stromal tumors.

catheter

A tube-shaped device that is used to give treatment or drain fluid from the body.

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.

clinical trial

A type of research that assesses health tests or treatments.

colonoscopy

A procedure to work inside the colon with a device that is guided through the anus.

computed tomography (CT)

A test that uses x-rays from many angles to make a picture of the insides of the body.

contrast

A dye 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.

deoxyribonucleic acid (DNA)

A chain of chemicals in cells that contains coded instructions for making and controlling cells. Also called the “blueprint of life.”

desmoid tumor

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

embolization

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

endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA)

A procedure that removes fluid with a needle on an imaging device guided through a natural opening.

external beam radiation therapy (EBRT)

A cancer treatment with radiation delivered from a machine outside the body.

familial adenomatous polyposis (FAP)

A health condition that is passed down in a family and increases the chance of getting sarcoma.

fascia

A deep layer of soft tissue.

fine-needle aspiration (FNA)

A procedure that removes tissue samples with a very thin needle.

Gardner's syndrome

A health condition that is passed down in families and increases the chance of getting sarcoma.

gastroenterologist

A doctor who's an expert in digestive diseases.

gene

Coded instructions in cells for making new cells and controlling how cells behave.

general anesthesia

A drug-induced, sleep-like state for pain relief.

genetic assessment

A lab test of abnormal coded instructions in cells that are passed down within a family.

hereditary

Passed down from parent to child through coded information in cells.

hives

A skin rash caused by the body trying to rid itself of a foreign substance.

imaging

A test that makes pictures (images) of the insides of the body.

immune system

The body's natural defense against infection and disease.

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.

intraoperative radiation therapy (IORT)

Radiation therapy given during surgery.

isolated limb infusion/perfusion

A method of giving cancer drugs into the controlled bloodstream of a limb or arm.

KIT

A protein on the edge of a cell that send signals for the cell to grow.

laparoscopic surgery

An operation with tools that are passed through small cuts in the belly area.

Li-Fraumeni syndrome

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

lymph node

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

magnetic resonance imaging (MRI)

A test that uses radio waves and powerful magnets to make pictures of the insides of the body.

medical history

A report of all your health events and medications.

medical oncologist

A doctor who's an expert in cancer drugs.

metastasectomy

Surgery to remove cancer that has spread far from the first tumor.

metastasis

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

mutation

An abnormal change.

neoadjuvant treatment

A treatment that is given before the main treatment to reduce the cancer. Also called preoperative treatment if given before an operation.

nutritionist

A health care worker who completed education in food and diet.

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.

pediatric oncologist

A doctor who's an expert in treating cancer in children.

pelvis

The body area between the hipbones.

physical exam

A study of the body by a health expert for signs of 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.

platelet-derived growth factor receptor alpha (PDGFRA)

A protein on the edge of a cell that sends signals for the cell to grow.

positron emission tomography (PET)

A test that uses radioactive material to see the shape and function of body parts.

primary treatment

The main treatment used to rid the body of cancer.

primary tumor

The first mass of cancer cells.

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 treatment that uses high-energy rays or related approaches to kill cancer cells.

radiologist

A doctor who's an expert in imaging tests.

retroperitoneum

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

sarcoma

A cancer of bone or soft tissue cells.

sedative

A drug that helps a person to relax or go to sleep.

side effect

An unhealthy or unpleasant physical or emotional response to treatment.

simulation

The steps needed to prepare for treatment with radiation.

social worker

An expert in meeting social and emotional needs.

soft tissue sarcoma

A cancer of cells that support, connect, and surround parts of your body.

stereotactic body radiation therapy (SBRT)

Treatment with high-dose radiation within one or a few sessions. Also called stereotactic ablative radiotherapy (SABR).

succinate dehydrogenase (SDH)

A protein within cells.

supportive care

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

surgery

An operation to remove or repair a part of the body.

surgical margin

The normal-looking tissue around the edge of a tumor that is 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.

three-dimensional conformal radiation therapy (3D-CRT)

A treatment with radiation that uses beams matched to the shape of the tumor.

TP53

An abnormal change in cells' coded instructions (genes) that causes Li-Fraumeni syndrome.

ultrasound

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

x-ray

A test that uses small amounts of radiation to make pictures of the insides of the body. Also called a plain radiograph.

Acronyms

3D-CRT

three-dimensional conformal radiation therapy

AJCC

American Joint Commission on Cancer

APC

adenomatous polyposis coli

CAM

complementary and alternative medicine

CBC

complete blood count

CNS

central nervous system

CT

computed tomography

DNA

deoxyribonucleic acid

EBRT

external beam radiation therapy

EUS-FNA

endoscopic ultrasound-guided fine-needle aspiration

FAP

familial adenomatous polyposis

FNA

fine-needle aspiration

FNCLCC

French Federation of Cancer Centers Sarcoma Group

GIST

gastrointestinal stromal tumor

HDR brachytherapy

high dose-rate brachytherapy

IMRT

intensity-modulated radiation therapy

IORT

intraoperative radiation therapy

LDR brachytherapy

low dose-rate brachytherapy

MRI

magnetic resonance imaging

NCI

National Cancer Institute

NSAID

nonsteroidal anti-inflammatory drug

PDGFRA

platelet-derived growth factor receptor alpha

PET

positron emission tomography

PET/CT

positron emission tomography/computed tomography

RMS

rhabdomyosarcoma

SBRT

stereotactic body radiation therapy

SDH

succinate dehydrogenase

TNM

tumor, node, metastasis

TKI

tyrosine kinase inhibitor

Types of sarcoma

Histopathologic type: Tumors included in the soft tissue category are listed below as per the 2013 World Health Organization classification of tumors:

Adipocytic Tumors

Atypical lipomatous tumor
Well-differentiated liposarcoma
Liposarcoma, NOS
Dedifferentiated liposarcoma
Myxoid/round cell liposarcoma
Pleomorphic liposarcoma

Fibroblastic/Myofibroblastic Tumors

Dermatofibrosarcoma protuberans
Fibrosarcomatous dermatofibrosarcoma protuberans
Pigmented dermatofibrosarcoma protuberans
Solitary fibrous tumor, malignant
Inflammatory myofibroblastic tumor
Low-grade myofibroblastic sarcoma
Adult fibrosarcoma
Myxofibrosarcoma (formerly myxoid malignant fibrous histiocytoma [myxoid MFH])
Low-grade fibromyxoid sarcoma
Sclerosing epithelioid fibrosarcoma

So-called Fibrohistiocytic Tumors

Giant cell tumor of soft tissues

Smooth Muscle Tumors

Leiomyosarcoma (excluding skin)

Pericytic (Perivascular) Tumors

Malignant glomus tumor

Skeletal Muscle Tumors

Embryonal rhabdomyosarcoma (including botryoid, anaplastic)
Alveolar rhabdomyosarcoma (including solid, anaplastic)
Pleomorphic rhabdomyosarcoma
Spindle cell/sclerosing rhabdomyosarcoma

Vascular Tumors

Retiform hemangioendothelioma
Pseudomyogenic (epithelioid sarcoma-like) hemangioendothelioma
Epithelioid hemangioendothelioma
Angiosarcoma of soft tissue

Chondro-osseous Tumors

Extraskeletal osteosarcoma

Gastrointestinal Stromal Tumors

Gastrointestinal stromal tumor, malignant

Nerve Sheath Tumors

Malignant peripheral nerve sheath tumor
Epithelioid malignant peripheral nerve sheath tumor
Malignant Triton tumor
Malignant granular cell tumor

Tumors of Uncertain Differentiation

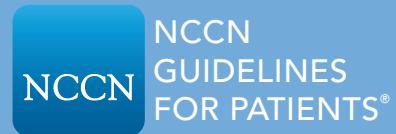
Ossifying fibromyxoid tumor, malignant
Stromal sarcoma, NOS
Myoepithelial carcinoma
Phosphaturic mesenchymal tumor, malignant
Synovial sarcoma (NOS, spindle cell, biphasic)
Epithelioid sarcoma
Alveolar soft part sarcoma
Clear cell sarcoma of soft tissue
Extraskeletal myxoid chondrosarcoma
Extraskeletal Ewing sarcoma
Desmoplastic small round cell tumor
Extrarenal rhabdoid tumor
Perivascular epithelioid cell tumor (PEComa), NOS
Intimal sarcoma

Undifferentiated/Unclassified Sarcoma

Undifferentiated (spindle cell sarcoma, pleomorphic sarcoma, round cell sarcoma, epithelioid sarcoma, NOS)

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Need a great resource on cancer?



Let us be your guide

NCCN Guidelines for Patients® empower you to make informed decisions at each step of your cancer journey.

CANCER TREATMENT:

- Acute Lymphoblastic Leukemia
- Acute Myeloid Leukemia
- Brain Cancer/Gliomas
- Breast Cancer
 - Noninvasive Breast Cancer
 - Early-Stage (Stages I and II)
 - Locally Advanced (Stage III)
 - Metastatic Breast Cancer
- Chronic Lymphocytic Leukemia
- Chronic Myeloid Leukemia
- Colon Cancer
- Esophageal Cancer
- Hepatobiliary Cancers
 - Liver, Gallbladder, and Bile Duct Cancers
- Hodgkin Lymphoma
- Kidney Cancer
- Lung Cancer
 - Non-Small Cell Lung Cancer

- Malignant Pleural Mesothelioma
- Melanoma
- Multiple Myeloma
- Myelodysplastic Syndromes
- Myeloproliferative Neoplasms
- Non-Hodgkin's Lymphomas
 - Diffuse Large B-cell Lymphoma
 - Follicular Lymphoma
 - Mantle Cell Lymphoma
 - Mycosis Fungoides
 - Peripheral T-cell Lymphoma
- Ovarian Cancer
- Pancreatic Cancer
- Prostate Cancer
- Rectal Cancer
- Soft Tissue Sarcoma
- Stomach Cancer
- Thyroid Cancer
- Waldenström's Macroglobulinemia/ Lymphoplasmacytic Lymphoma

SUPPORTIVE CARE:

- Distress
- Nausea and Vomiting

CANCER SCREENING:

- Lung Cancer Screening

AGE-RELATED:

- Adolescents and Young Adults (AYAs) with Cancer

TRANSLATIONS:

- Kidney Cancer
 - Chinese
 - Czech
 - German
 - Spanish
- Stomach Cancer
 - Italian



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NCCN.org/patients



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206.667.5000 • fredhutch.org

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hopkinskimmelcancercenter.org

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866.587.4322
cancer.northwestern.edu

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904.953.0853 • Florida
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mskcc.org

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moffitt.org

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

Roswell Park Comprehensive
Cancer Center
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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
888.226.4343 • stjude.org
901.683.0055 • westclinic.com

Stanford Cancer Institute
Stanford, California
877.668.7535
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www3.ccc.uab.edu

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vicc.org

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855.4.SMILOW
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Soft Tissue Sarcoma

2018

NCCN Foundation® gratefully acknowledges our advocacy supporters SARC (Sarcoma Alliance for Research through Collaboration) and Rockin for the Cure, and our industry supporter Eisai Inc. for their support in making available these NCCN Guidelines for Patients®. NCCN independently develops and distributes the NCCN Guidelines for Patients. Our industry supporters do not participate in the development of the NCCN Guidelines for Patients and are not responsible for the content and recommendations contained therein.



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