

# Soft Tissue Sarcoma







# Soft Tissue Sarcoma

Learning that you have soft tissue sarcoma can be overwhelming. The goal of this book is to help you get the best care. It presents 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 25 of the world's leading cancer centers. Experts from NCCN® have written treatment guidelines for doctors who treat colon cancer. 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. NCCN also offers patient books on lung cancer, melanoma, and many other cancer types. Visit [NCCN.org/patients](http://NCCN.org/patients) for the full library of patient books as well as other resources.

# Credits

NCCN aims to improve the care given to patients with cancer. NCCN staff work with experts to create helpful programs and resources for many stakeholders. Stakeholders include health providers, patients, businesses, and others. One resource is the series of books for patients called the NCCN Patient Guidelines®. Each book presents the best practice for a type of cancer.

The patient books are based on clinical practice guidelines written for cancer doctors. These guidelines are called the NCCN Guidelines®. Clinical practice guidelines list the best health care options for groups of patients. Many doctors use them to help plan cancer treatment for their patients.

Panels of experts create the NCCN Guidelines. Most of the experts are from the 25 NCCN Member Institutions. Panelists may include surgeons, radiation oncologists, medical oncologists, and patient advocates. 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](http://NCCN.org/clinical.asp).

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The NCCN Foundation supports the mission of the National Comprehensive Cancer Network® (NCCN®) to improve the care of patients with cancer. One of its aims is to raise funds to create a library of books for patients. Learn more about the NCCN Foundation at [NCCN.org/foundation](http://NCCN.org/foundation).

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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.

## Does the whole book apply to me?

Part 1 briefly describes sarcoma and its treatments. Parts 2 through 5 offer a treatment guide for different types of tumors. Part 6 gives tips for anyone making treatment decisions.

This book includes information for many situations. However, it doesn't address treatment for rhabdomyosarcoma, Ewing's sarcoma, and desmoplastic small round cell tumor. Part 2 covers treatment for sarcomas in the arms, legs, trunk wall, head, or neck. Part 3 covers sarcomas in the space in front of your lower spine (retroperitoneum), belly area (abdomen), and between the hip bones (pelvis). Part 4 is a treatment guide for GISTs (**g**astrointestinal **s**tromal **t**umors). The last treatment guide, Part 5, discusses desmoid tumors.

Your treatment team can help. They can point out what information applies 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.

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

## Making sense of medical terms

In this book, many medical words are included that describe cancer, tests, and treatments. These are words that 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. Don't be shy to ask your treatment team to explain a word or phrase that you do not understand.

Words that you may not know are defined in the text or in the *Dictionary*. Words in the *Dictionary* are underlined when first used on a page.

Acronyms are also defined when first used and in the *Glossary*. Acronyms are short words formed from the first letters of several words. One example is FAP for **f**amilial **a**denomatous **p**olyposis.

# Sarcoma basics





# 1 Sarcoma basics

6	What is soft tissue sarcoma?
8	Local treatments
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Learning that you have cancer can be overwhelming. This chapter briefly describes what sarcoma is and the types of treatment. These basics may help you cope and better understand Parts 2 through 6.

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## What is 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.

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 can be found in the *Glossary*. The most common types are:

- Undifferentiated pleomorphic sarcoma,
- GISTs (**g**astrointestinal **s**tral tumors),
- Liposarcoma,
- Leiomyosarcoma,
- Synovial sarcoma, and
- Malignant peripheral nerve sheath tumors.

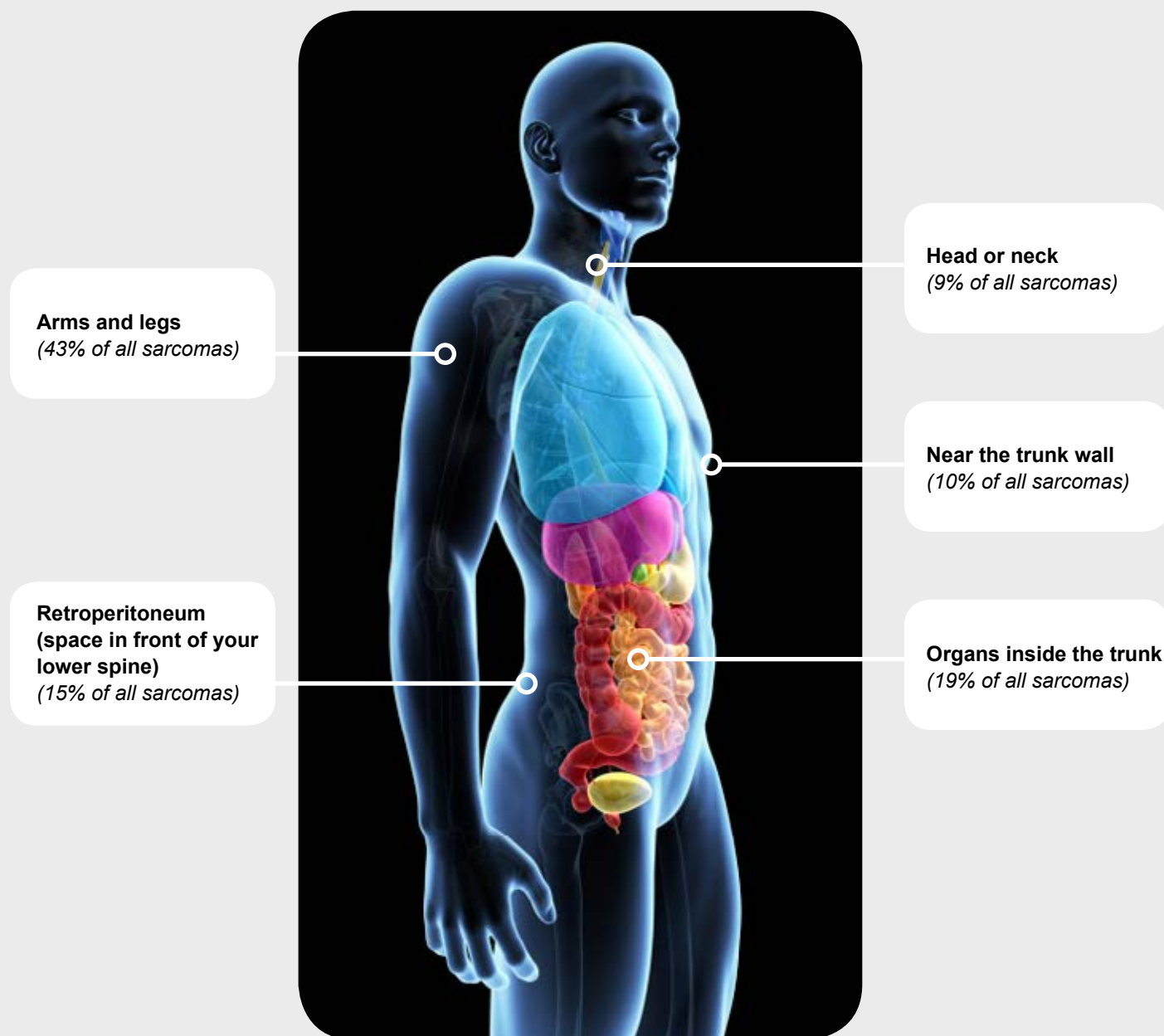
Soft tissue sarcomas can occur anywhere in the body. The most common place is in the arms and legs. About 43 out of 100 (43%) sarcomas occur in the



limbs. The second most common site is organs inside the trunk, such as the stomach and intestines. About 19% of sarcomas occur here. Next, 15% of sarcomas occur here. Next, 15% of sarcomas

occur in a space in front of your lower spine called the retroperitoneum. About 10% of sarcomas occur in or near the trunk wall and 9% occur in the head or neck.

Figure 1.1 Most common locations for sarcoma in the body.



## Local treatments

Local treatments are used to treat a focused area of cancer. Surgery and radiation therapy are common local treatments. Ablation and embolization are sometimes used for small tumors that aren't near to where the cancer started. One example is small tumors in the liver.

### Surgery

Surgery has been used as a primary treatment for sarcoma for a long time. Primary treatment is the main treatment used to rid the body of cancer. As such, 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 remove a cancer-free surgical margin. A surgical margin is a ring of normal-looking tissue around the tumor. Sometimes a cancer-free margin can't be removed. If this is expected, your surgeon will place clips in your body after removing the tumor. The clips help doctors give you radiation therapy where it is needed. Sometimes, a second surgery can be done instead.

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.

Side effects are unhealthy or unpleasant physical or emotional responses to treatment. You may experience side effects from the anesthesia or the surgery. Often, general anesthesia is used for

surgery. General anesthesia is drugs that put you into a deep sleep-like state so you won't feel pain. Ask your treatment team for a list of all rare and common side effects of the surgery you will have.

### Radiation therapy

Radiation therapy uses high-energy rays to treat cancer. The rays damage DNA (deoxyribonucleic acid). DNA is a chain of chemicals in cells that contains genes. This either kills the cancer cells or stops new cancer cells from being made.

Radiation therapy has many uses for sarcomas. It may be used as a primary treatment like surgery. Radiation therapy given before surgery is called neoadjuvant treatment. It is used to shrink the tumor for surgery. Radiation therapy can also be given during or after surgery to kill any remaining cancer cells. Radiation therapy given during surgery is called IORT (intraoperative radiation therapy). When given after surgery, it is called adjuvant treatment. You may receive a full dose of radiation, or if receiving a "boost," less than a full dose. A boost is only used for neoadjuvant treatment.

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

### External beam radiation therapy

Radiation is often given using a machine outside the body. This method is called EBRT (external beam radiation therapy). 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 shape of radiation

beams, and number of treatment sessions. Beams are shaped with computer software and hardware added to the radiation machine. Radiation beams are aimed at the tumor with help from ink marks on the skin 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. For sarcoma, 3D-CRT (**three-dimensional conformal radiation therapy**), IMRT (**intensity-modulated radiation therapy**), or SBRT (**stereotactic body radiation therapy**) may be used. In 3D-CRT, the radiation beams match the shape of the tumor to avoid healthy tissues. IMRT is a more precise type of 3D-CRT. The radiation beam is divided into smaller beams, and the strength of each beam can vary. 3D-CRT and IMRT take several weeks to complete. SBRT is completed in 1 to 2 weeks and treats sarcomas with precise but high-dose beams.

3D-CRT and IMRT use photon radiation beams. Photon beams are a stream of particles that have no mass or electric charge. In recent years, a few cancer centers have built radiation machines that use proton beams. Proton beams are a stream of positively charged particles that emit energy within a short distance. SBRT uses either photons or protons.

### 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 (**low-dose rate**) or HDR (**high-dose rate**).

For LDR brachytherapy, the radioactive objects will remain in your body for a few days. The objects give off low doses of radiation during the whole time the objects are in you. The radiation travels a very short distance.

HDR brachytherapy is a newer method. It gives more radiation than LDR, but the radiation stays in your body for a shorter period of time. Before treatment, imaging will be done to assure the catheters are in place. A machine will then pump the radioactive source into the catheters. After treatment, the machine will remove the radioactive source from your body. You may receive HDR more than once a day for a short period of time. HDR brachytherapy can also be used as IORT.

### 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 (**computed tomography**) 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.

## Drug treatments

Drugs are another type of treatment for cancer. Drugs can travel in blood and reach cancer cells anywhere in the body. Chemotherapy, targeted therapy, and immunotherapy are cancer drugs used for sarcomas. [See Chart 1.1](#) for a list of drug names recommended by NCCN. Which drugs are used depends on the type of sarcoma.

### Chemotherapy

Chemotherapy, or ‘chemo,’ is a class of drugs that is used to kill cancer cells. Some chemotherapy drugs kill cancer cells by damaging their DNA or disrupting the making of DNA. Other drugs interfere with cell parts that are needed for making new cells.

Chemotherapy has many uses. It is used as neoadjuvant, primary, or adjuvant treatment. It is also used to treat metastases.

Most chemotherapy drugs for sarcoma are liquids that are injected into a vein. Others are pills. Chemotherapy drugs differ in the way they work, so often more than one drug is used. A combination regimen is the use of two or more chemotherapy drugs.

Chemotherapy is often given in cycles of treatment days followed by days of rest. The cycles vary in length depending on which drugs are used. Giving chemotherapy in cycles gives your body a chance to recover after receiving chemotherapy. If you will have chemotherapy, ask your doctor if the chemotherapy will be given in cycles. If it will be then ask how many cycles and days of treatment there will be.

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.

In general, side effects are caused by the death of fast-growing normal cells. These cells are found in the hair follicles, gut, mouth, and blood. Thus, 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 known common and rare side effects.

### Chemoradiation

Chemotherapy and radiation given together is called chemoradiation. Sometimes these treatments are given at the same time. Sometimes, the treatments are staggered. For example, chemotherapy is given, then radiation, and then more chemotherapy.

### 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.

### 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.

Chart 1.1 Drug treatments for sarcoma

Generic (chemical) name	Brand name (sold as)	Type of drug
Bevacizumab	Avastin®	Targeted therapy
Celecoxib	Celebrex®	Targeted therapy
Crizotinib	Xalkori®	Targeted therapy
Dacarbazine	DTIC-Dome®	Chemotherapy
Dasatinib	Sprycel®	Targeted therapy
Docetaxel	Taxotere®	Chemotherapy
Doxorubicin	–	Chemotherapy
Doxorubicin hydrochloride, injection	Doxil®	Chemotherapy
Epirubicin hydrochloride	Ellence®	Chemotherapy
Everolimus	Afinitor®	Chemotherapy
Gemcitabine hydrochloride	Gemzar®	Chemotherapy
Ifosfamide	–	Chemotherapy
Imatinib mesylate	Gleevec®	Targeted therapy
Interferon alfa 2b, recombinant	Intron® A	Immunotherapy
Mesna	Mesnex® Injection	Chemotherapy
Methotrexate	–	Chemotherapy
Nilotinib	Tasigna®	Targeted therapy
Paclitaxel	Taxol®	Chemotherapy
Pazopanib	Votrient®	Targeted therapy
Regorafenib	Stivarga®	Targeted therapy
Sorafenib tosylate	Nexavar®	Targeted therapy
Sirolimus	Rapamune®	Chemotherapy
Sulindac	Clinoril®	Targeted therapy
Sunitinib malate	Sutent®	Targeted therapy
Tamoxifen citrate	–	Targeted therapy
Temozolomide	Temodar®	Chemotherapy
Toremifene citrate	Fareston®	Targeted therapy
Vinblastine sulfate	–	Chemotherapy
Vinorelbine tartrate	Navelbine®	Chemotherapy

## Clinical trials

New tests and treatments aren't offered to the public as soon as they're made. They need to be studied. New uses of tests and treatments also need to be studied.

A clinical trial is a type of research that studies a test or treatment. Clinical trials study how safe and helpful tests and treatments are. When found to be safe and helpful, they may become tomorrow's standard of care. Because of clinical trials, the tests and treatments in this book are now widely used to help patients.

Tests and treatments go through a series of clinical trials to make sure they're safe and work. Without clinical trials, there is no way to know if a test or treatment is safe or helpful. Clinical trials have four phases. Examples of the four phases for treatment are:

- **Phase I trials** aim to find the best dose of a new drug with the fewest side effects.
- **Phase II trials** assess if a drug works for a specific type of cancer.
- **Phase III trials** compare a new drug to the standard treatment.
- **Phase IV trials** involve drugs already approved by the U.S. FDA (**F**ood and **D**rug **A**dministration) for at least one disease.

Joining a clinical trial has benefits. First, you'll have access to the most current cancer care. Second, you will receive the best management of care. Third, the results of your treatment—both good and bad—will be carefully tracked. Fourth, you may help other patients with cancer.

Clinical trials have risks, too. Like any test or treatment, there may be side effects. Also, new tests or treatments may not work better than current treatments. Another downside may be that there may be more paperwork or more trips to the hospital.

To join a clinical trial, you must meet the conditions of the study. Patients in a clinical trial are often alike in terms of their cancer and general health. This is to know that any progress seen at the end of the study is because of the treatment and not because of differences between patients. To join, you'll need to review and sign a paper called an informed consent form. This form describes the study in detail, including the risks and benefits.

Ask your treatment team if there is an open clinical trial that you can join. 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 6.

## Review

- Soft tissue supports, connects, and surrounds parts of your body.
- Local treatments for soft tissue sarcoma include surgery, radiation therapy, ablation, and embolization.
- Drug treatments include chemotherapy, targeted therapy, and immunotherapy.
- Clinical trials give people access to new tests and treatments.

# 2

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





## 2 Sarcomas in limbs, outer trunk, head, or neck

14	Treatment planning
17	Treatment by stage
18	Stage I sarcoma
20	Stages II and III sarcoma
24	Stage IV sarcoma
26	Recurrence treatment
28	Review



This chapter is about sarcomas in the arms, legs, trunk wall, 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.

### Treatment planning

#### Must haves

Multidisciplinary team
Medical history and physical exam
Imaging of tumor
Biopsy
Imaging of chest

#### Sometimes useful

PET (positron emission tomography) of tumor
CT (computed tomography) of abdomen and pelvis
MRI (magnetic resonance imaging) of total spine
Imaging of central nervous system
Genetic assessment

### Multidisciplinary team

Treatment of sarcoma takes a team of experts who have experience with this cancer. If you have sarcoma, it is important that all the experts meet before your treatment is started to create the best treatment plan. Your treatment team will also meet while you are going through treatment and afterward to discuss the treatment results and the next steps of care. Your team of experts may include a:

In most cases

- Pathologist—an expert in testing cells and tissue to find 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, and a
- Nurse—an expert trained to care for the sick.

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 live life unaided or with devices,
- Physical therapist—an expert in helping people move better,
- Nutritionist—an expert in healthy foods and drinks, and a
- Genetic counselor—an expert in explaining testing for hereditary diseases.

### Medical history and physical exam

Your medical history includes any health events in your life. It also includes any medications you've taken or are taking. Since some health problems run in families, your doctor will ask about the medical history of your blood relatives.

Sarcoma often occurs for unknown reasons. But some people have syndromes that increase their chances of getting sarcoma. Li-Fraumeni syndrome is one such syndrome. It can be passed down from parents to child (inherited) or caused by other factors. It is very rare. Your doctors will assess if you likely have Li-Fraumeni syndrome.

Doctors often perform a physical exam along with taking a medical history. A physical exam is a review of your body for signs of disease. During this exam, your doctor will listen to your lungs, heart, and gut. Parts of your body will likely be felt to see if organs are of normal size, are soft or hard, or cause pain when touched. Your lymph nodes may feel large if cancer has spread to them.

### Imaging of tumor

Imaging tests make pictures (images) of the insides of your body. If a tumor is likely cancer, you should get an imaging scan of the tumor. You may receive an MRI with or without CT. MRI uses radio waves and powerful magnets to make images. CT takes many x-rays of the same body part from different angles to make detailed images. These tests can show your doctors how large a tumor is and how close it is to other tissues. For some people, less detailed imaging scans, such as an angiogram or plain radiograph, may be enough.

A contrast dye may be used to make the images clearer. The dye will be injected into your vein, mixed with a liquid you drink, or both. The dye may cause you to feel flushed or get hives. Rarely, serious allergic reactions occur. Tell your doctor and the technicians if you have had bad reactions in the past.

Getting an imaging scan is often easy. Before the scan, you may need to stop taking some medicines, stop eating and drinking for a few hours, and remove metal objects from your body. During the scan, you will need to lie face up on a table that moves through the machine. As the machine takes pictures, you may hear buzzing, clicking, or whirring sounds. You will be alone, but a technician will operate the machine in a nearby room. He or she will be able to see, hear, and speak with you at all times. You will likely be able to resume your activities right away unless you took a sedative.

## Biopsy

A biopsy is the removal of tissue or fluid samples to test for disease. A biopsy is recommended after adequate imaging of the tumor. After the biopsy, a pathologist will examine the samples with a microscope to confirm if there's cancer.

A biopsy can be done with a needle. There are two types of needle biopsies. A core-needle biopsy is the more common and favored method. It obtains a larger sample for testing. A fine-needle aspiration removes a smaller piece of tissue and doesn't require any cutting into the skin (incision).

Besides needle biopsies, an incisional biopsy can be used. An incisional biopsy requires anesthesia to numb the area and a cut into the skin. It is a minor surgery that removes some but not the entire tumor.

If after the first biopsy, it is unclear if the tumor is cancer, a second biopsy should be done. The second biopsy should be done using imaging to guide the needle into the tumor. If the tumor is cancer, more testing should be done to learn more about the cancer. Your doctors will want to know the cancer grade, which is discussed on page 17. Testing for abnormal genes within the cancer cells may also be done. It is important that the testing be done by a pathologist who has experience with sarcoma.

## Imaging of chest

Chest imaging is recommended before treatment of sarcomas that are discussed in this chapter. Imaging of your chest will allow your doctors to assess if the cancer has spread to your lungs. You may receive a plain radiograph or CT of the chest. Chest CTs are often used when images from a plain x-ray would be unclear due to lung scarring or other health conditions.

## PET

PET scans are a type of imaging test. To create the images, a sugar radiotracer is used. If the radiotracer is FDG (fluorodeoxyglucose), you must fast for 4 hours before it is injected into your body. Between 40 to 60 minutes after injection, the radiotracer will emit a small amount of energy. This energy can be detected by the imaging machine.

In the PET images, cancer appears brighter ("hotter"). This is because cancer cells use sugar more quickly than normal cells. Hot spots suggest that cancer is present.

PET may be useful if the tumor is larger than 3 cm, is firm, and is deep within your body. PET may help your doctors decide the prognosis and grade of the cancer. It may also help them assess if a tumor is shrinking during chemotherapy.

## CT of abdomen and pelvis

Some sarcomas are more likely to spread to other sites than other sarcomas. These include myxoid and round cell liposarcoma, epithelioid sarcoma, angiosarcoma, and leiomyosarcoma. If you have these sarcomas, you may receive a CT scan of your abdomen and pelvis to assess if the cancer has spread. CT of these areas is also done if this is where the sarcoma started. In this case, CT may show if tumor has come back or spread to the organs in the abdomen.

## MRI of total spine

Myxoid and round cell liposarcomas are likely to

spread to the bones. Examples of bones include the spine and pelvis. If you have these sarcomas, you may receive an MRI of total spine to assess if the cancer has spread.

### Imaging of central nervous system

Alveolar soft-part sarcomas and angiosarcomas are more likely to spread to the brain. This is especially true for stage IV alveolar soft part sarcomas that have spread to the lungs. If you have these sarcomas, you may receive imaging scans of your central nervous system. The central nervous system includes your brain and spinal cord.

### Genetic assessment

If you may have Li-Fraumeni syndrome, your doctor may want you to have a genetic assessment. A genetics counselor will explain what the goals, benefits, and risks of the assessment are. The counselor may also ask more questions about your health and your family's health history. You may decide to get tested for Li-Fraumeni syndrome. To be tested, you must provide a sample of body tissue. Using the tissue sample, a pathologist can test your genes for the TP53 mutations that cause Li-Fraumeni syndrome.

## 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 (**A**merican **J**oint **C**ommittee on **C**ancer) staging system is used to stage sarcoma. There are four stages—I, II, III, and IV.

In this system, the letters T, N, and M describe a different area of cancer growth. The T score describes the growth of the primary tumor. The N score describes spread of cancer growth to lymph nodes. The M score tells if the cancer has spread to

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

Another factor used in staging is the cancer grade. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. A three-grade system is used for sarcoma based on scores of:

- how much the cancer cells look like normal cells,
- how fast the cancer cells are making copies of themselves, and
- how much dead tissue is in the tumor.

The stages of sarcoma are:

### Stage I

- Stage IA tumors are 5 cm or smaller (T1). They can be superficial (T1a) or deep (T1b). The cancer has not spread to lymph nodes (N0) or more distant sites (M0). Stage IA tumors are low grade (G1) or the grade is unknown (GX).
- Stage IB is the same as stage IA except the primary tumor is larger than 5 cm (T2).

### Stage II

- Stage IIA tumors are 5 cm or smaller (T1), have not spread (N0, M0), and are either intermediate (G2) or high (G3) grade.
- Stage IIB tumors are larger than 5 cm, have not spread, and are intermediate grade (G2).

### Stage III

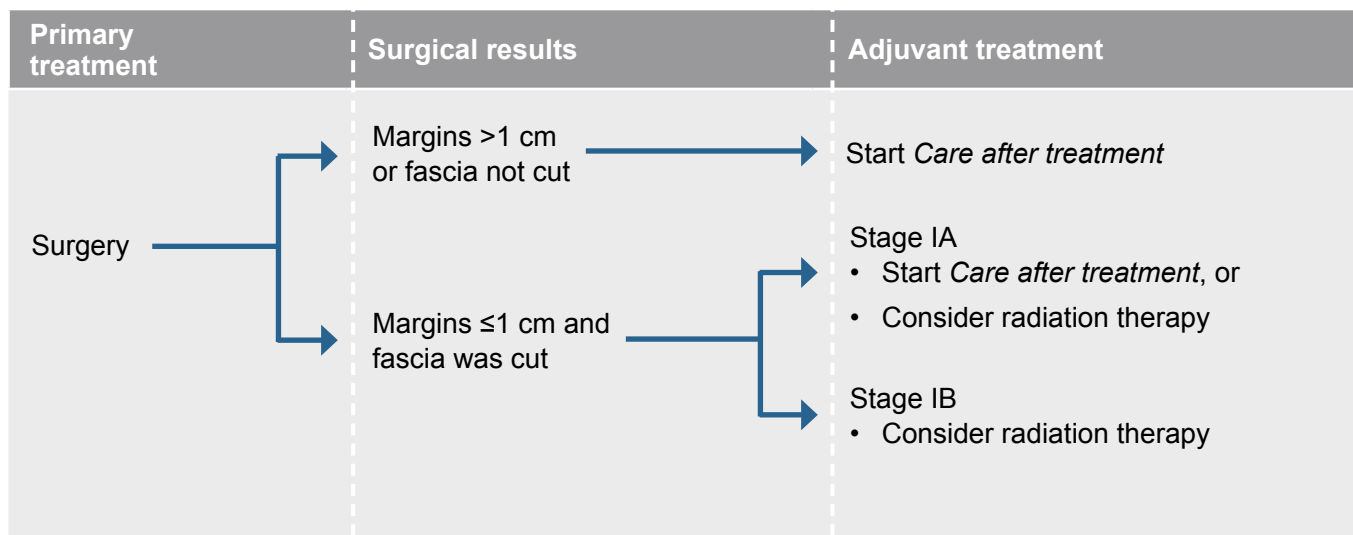
- The primary tumor is larger than 5 cm (T2), has not spread (N0, M0), and is high grade (G3). Stage III tumors can also be of any size or grade and spread to lymph nodes (N1, M0).

### Stage IV

- These sarcomas can be of any size or grade and have spread to distant sites (M1).

## 2.1 Stage I sarcoma

**Chart 2.1.1 Treatment options**



**Chart 2.1.2 Care after treatment**

Type of care	Schedule of care
• Rehabilitation	If needed
• Medical history and physical exam	Every 3–6 months for 2–3 years, <ul style="list-style-type: none"> <li>◦ If good results, then repeat every year</li> </ul>
• Consider chest imaging	Every 6–12 months
• Consider imaging of tumor site	Right after surgery then at regular intervals of time

**Chart 2.1.1** maps the treatment options for stage I sarcoma. Surgery to remove the tumor is recommended for primary treatment. For stage I, surgery is done to cure the cancer.

Removing 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.

After surgery, your doctors will assess if you need adjuvant treatment. Adjuvant treatment can help stop the cancer from returning. 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.

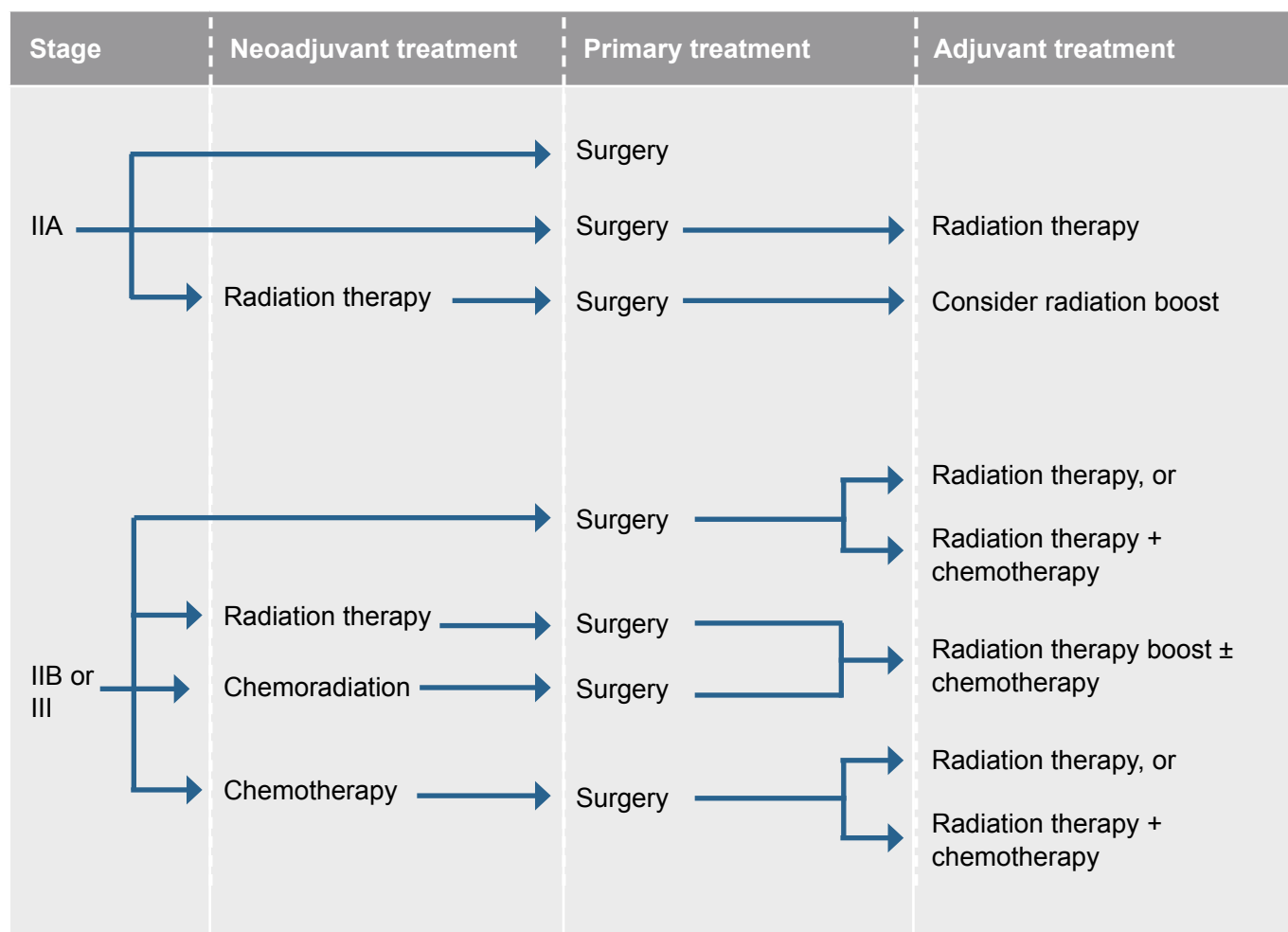
No more treatment will be needed if the surgical margin is larger than 1 cm or the fascia was not cut. Treatment options are based on cancer stage if the surgical margin is 1 cm or smaller and the fascia was cut. For stage IA, the first option is no further treatment. For some stage IA cancers, your doctor may consider radiation therapy. Radiation therapy may lower the chances of the cancer returning. For stage IB, radiation therapy should be considered.

**Chart 2.1.2** lists care after cancer treatment has ended. You should receive rehabilitation if you need it. This may include occupational or physical therapy. You should also start to have follow-up tests to check if the cancer has returned. Getting follow-up tests can help find cancer early. Cancer is more likely to be cured if found early.

Tests include 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.

## 2.2 Stages II and III sarcoma

**Chart 2.2.1 Surgery is an option**



Treatment options for sarcoma are based on whether surgery can be done. Some sarcomas can't be removed by surgery at first 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 can be done are listed next followed by options when surgery may not be possible.

**Chart 2.2.1** maps the treatment options for stage II and III sarcomas that can be treated with surgery. For stage IIA, surgery alone may be an option. Surgery alone may be done if the tumor is small and the surgical margins can be wide.

The second option for stage IIA 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.

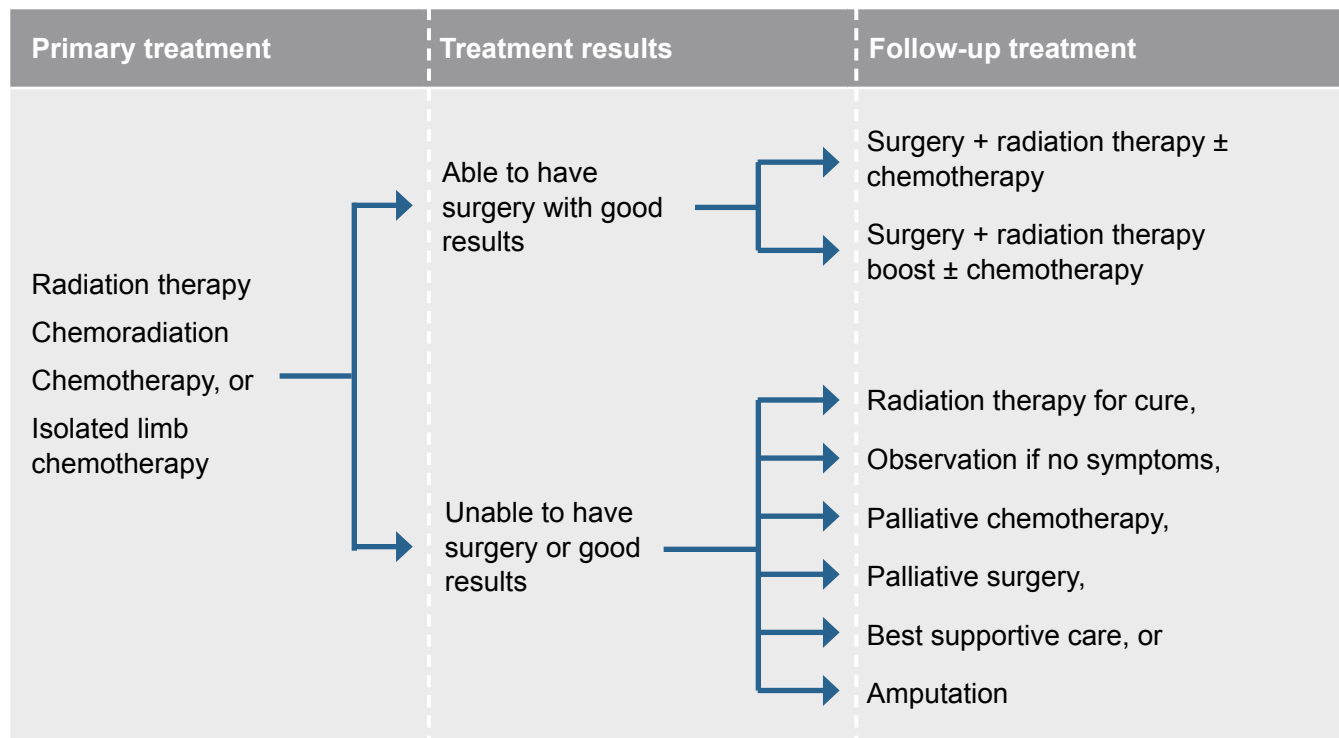


The third option for stage IIA is to have radiation therapy followed by surgery. Radiation therapy before surgery may reduce the chances of the cancer returning. It may also improve how well your limb works after surgery. However, radiation will likely slow healing of the surgical wound. Before having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

There are four options for stages IIB and III. All four include surgery to remove the tumor. Your lymph nodes should also be removed if the cancer is stage III. The cancer has spread to the lymph nodes.

Surgery followed by radiation therapy is strongly recommended. Other options include radiation therapy, chemoradiation, or chemotherapy before surgery. A radiation boost may follow surgery that was preceded by radiation therapy. A boost may help if the cancer wasn't fully removed or cancer was found in the margins.

After surgery for IIB or III disease, you may receive chemotherapy. However, the research on its benefits at this point of care is limited. It is not clear if chemotherapy is helpful. Joining a clinical trial that is testing chemotherapy may be a good option.

**Chart 2.2.2 Surgery may not be an option****Chart 2.2.3 Care after treatment**

Type of care	Schedule of care
• Rehabilitation	If needed
• Medical history, physical exam, and chest imaging	Every 3–6 months for 2–3 years <ul style="list-style-type: none"> <li>◦ If good results, then repeat every 6 months for 2 years</li> <li>◦ If good results, then repeat every year</li> </ul>
• Consider imaging of tumor site	Right after surgery then at regular intervals of time

**Chart 2.2.2** maps the treatment options for stages II and III that can't be treated with surgery at first. However, other treatments may shrink the tumor so that surgery can be done. These treatments include radiation therapy, chemoradiation, chemotherapy, or isolated limb chemotherapy.

Isolated limb chemotherapy only treats the limb with sarcoma. The drugs are injected into blood vessels within the limb. This prevents chemotherapy from affecting the rest of your body.

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.

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

You may or may not also have chemotherapy after surgery. The research on its benefits at this point of care is limited. It is not clear if chemotherapy is helpful. Joining a clinical trial that is testing chemotherapy may be a good option.

There are treatment options if you are unable to have surgery. Amputation should be used as a last resort. Your doctors will consider many factors to decide which other options are best for you.

Some patients can have radiation therapy. The goal is to cure the cancer. The highest dose that doesn't cause severe side effects is recommended. If you will have EBRT, treatment planning should use IMRT, tomography, or protons.

If you can't have radiation therapy and 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.

Palliative care is recommended if the cancer is causing symptoms. There are three options. You may have chemotherapy, surgery, or best supportive care.

**Chart 2.2.3** 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. Getting follow-up tests can help find cancer early. Cancer is more likely to be cured if found early. Tests include medical history, physical exam, and imaging of your chest. If the cancer is likely to return, you may get imaging tests of the site where the primary tumor was.

## 2.3 Stage IV sarcoma

**Chart 2.3.1 Confined cancer spread**

Treatment options
<p>Treatment as listed for stages II and III on page 20 or consider these options:</p> <ul style="list-style-type: none"> <li>• Surgery to remove metastases ± chemotherapy, radiation therapy, or both before or after surgery,</li> <li>• Ablation,</li> <li>• Embolization,</li> <li>• SBRT, or</li> <li>• Observation</li> </ul>

**Chart 2.3.2 Care after treatment**

Type of care	Schedule of care
<ul style="list-style-type: none"> <li>• Rehabilitation</li> </ul>	If needed
<ul style="list-style-type: none"> <li>• Medical history, physical exam, chest imaging, and imaging of metastatic sites</li> </ul>	<p>Every 3–6 months for 2–3 years</p> <ul style="list-style-type: none"> <li>◦ If good results, then repeat every 6 months for 2 years</li> <li>◦ If good results, then repeat every year</li> </ul>
<ul style="list-style-type: none"> <li>• Consider imaging of tumor site</li> </ul>	Right after surgery then at regular intervals of time

**Chart 2.3.3 Widespread cancer**

Treatment options
<ul style="list-style-type: none"> <li>• Palliative chemotherapy</li> <li>• Palliative radiation therapy</li> <li>• Palliative surgery</li> <li>• Observation if no symptoms</li> <li>• Supportive care</li> <li>• Ablation</li> <li>• Embolization</li> <li>• SBRT</li> </ul>

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 don't join a clinical trial, the general recommendations presented next may help. General recommendations are divided into those for stage IV confined to one organ and widespread stage IV.

**Chart 2.3.1** 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 stage 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, radiation therapy, or both either before or after surgery. The other four options are ablation, embolization, SBRT, and observation.

**Chart 2.3.2** 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. Getting follow-up tests can help find cancer early. Cancer is more likely to be cured if found early. Tests include medical history, physical exam, and imaging of your chest. If the cancer is likely to return, you may get imaging tests of the site where the primary tumor was.

**Chart 2.3.3** lists the treatment options for widespread sarcoma. Treatment options depend on if the cancer is or isn't causing symptoms. 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 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.

## 2.4 Recurrence treatment

**Chart 2.4.1 Distant recurrences**

Cancer spread	Treatment options
Tumor in one organ and it's not large	<ul style="list-style-type: none"> <li>• Surgery to remove metastases ± chemotherapy, radiation, or both before or after surgery</li> <li>• Ablation</li> <li>• Embolization</li> <li>• SBRT</li> </ul>
Confined area	<ul style="list-style-type: none"> <li>• Lymph node surgery ± chemotherapy, radiation, or both</li> <li>• Surgery to remove metastases ± chemotherapy, radiation, or both before or after surgery</li> <li>• SBRT</li> </ul>
Widespread	<ul style="list-style-type: none"> <li>• Palliative chemotherapy</li> <li>• Palliative radiation therapy</li> <li>• Palliative surgery</li> <li>• Observation if no symptoms</li> <li>• Supportive care</li> <li>• Ablation</li> <li>• Embolization</li> <li>• SBRT</li> </ul>

Sarcoma may return as local disease. In this case, the cancer should be treated by its stage as described on pages 18 to 23. However, the return of sarcoma in distant sites should be treated as described below.

**Chart 2.4.1** lists the treatment options for a recurrence that is metastatic disease. Options are listed by the extent of the recurrence. First, 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, radiation therapy, or both

either before or after surgery. Other options include ablation, embolization, or SBRT.

The second list of treatment options is recurrences within a confined area. The cancer may be in lymph nodes that are near to where the cancer started. For confined disease, options include surgery to remove the lymph nodes with cancer. Surgery may also be done to remove other tumors. You may receive chemotherapy, radiation therapy, or both with the surgery. The last option is SBRT.

The third list of treatment options is for widespread sarcoma. Treatment options depend on if the cancer is or isn't causing symptoms. 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 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.



- Treatment planning is an important first step of care.
- Many 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.

- Clinical trials are the preferred treatment for stage IV sarcomas.
- Treatment options for recurrences are based on the extent of the cancer.

This image shows a blank sheet of white paper with horizontal ruling lines. The lines are evenly spaced and run across the width of the page. There are no margins, text, or other markings on the paper.

# Sarcomas in the inner trunk



# 3 Sarcomas in the inner trunk

30 Treatment planning

34 Treatment with surgery

36 Treatment without surgery

36 Review



This chapter is about sarcomas within the retroperitoneal, abdominal, or pelvic spaces. It starts by listing what is needed for treatment planning. Treatment options are then presented for when surgery can and can't be done.

## Treatment planning

### Must haves

Multidisciplinary team

Medical history and physical exam

Imaging of chest, abdomen, and pelvis

Biopsy if you:

- Will have neoadjuvant treatment,
- May have two types of cancer, or
- Won't be having surgery

### Sometimes useful

Biopsy if you will have surgery

Genetic assessment

### Multidisciplinary team

Treatment of sarcoma takes a team of experts who have experience with this cancer. If you have sarcoma, it is important that all the experts meet before your treatment is started to create the best treatment plan. Your treatment team will also meet while you are going through treatment and afterward to discuss the treatment results and the next steps of care. Your team of experts may include a:

In most cases

- Pathologist—an expert in testing cells and tissue to find 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, and a
- Nurse—an expert trained to care for the sick.

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 live life unaided or with devices,
- Physical therapist—an expert in helping people move better,
- Nutritionist—an expert in healthy foods and drinks, and a
- Genetic counselor—an expert in explaining testing for hereditary diseases.

### Medical history and physical exam

Your medical history includes any health events in your life. It also includes any medications you've taken or are taking. Since some health problems run in families, your doctor will ask about the medical history of your blood relatives.

Sarcoma often occurs for unknown reasons. But some people have syndromes that increase their chances of getting sarcoma. Li-Fraumeni syndrome is such a syndrome. It can be passed down from parents to child (inherited) or caused by other factors. It is very rare. Your doctors will assess if you likely have Li-Fraumeni syndrome.

Doctors often perform a physical exam along with taking a medical history. A physical exam is a review of your body for signs of disease. During this exam, your doctor will listen to your lungs, heart, and gut. Parts of your body will likely be felt to see if organs are of normal size, are soft or hard, or cause pain when touched. Your lymph nodes may feel large if cancer has spread to them.

### Imaging of chest, abdomen, and pelvis

Imaging tests make pictures (images) of the insides of your body. If a tumor is likely cancer, you should get an imaging scan of your chest, abdomen, and pelvis. You may receive CT with or without MRI. CT takes many x-rays of the same body part from different angles to make detailed images. MRI uses radio waves and powerful magnets to make images. These tests can show your doctors how large a tumor is and how close it is to other tissues. For some people, less detailed imaging scans, such as an angiogram or plain radiograph, may be enough.

A contrast dye should be used to make the images clearer. The dye will be injected into your vein, mixed with a liquid you drink, or both. The dye may cause you to feel flushed or get hives. Rarely, serious

allergic reactions occur. Tell your doctor and the technicians if you have had bad reactions in the past.

Getting an imaging scan is often easy. Before the scan, you may need to stop taking some medicines, stop eating and drinking for a few hours, and remove metal objects from your body. During the scan, you will need to lie face up on a table that moves through the machine. As the machine takes pictures, you may hear buzzing, clicking, or whirring sounds. You will be alone, but a technician will operate the machine in a nearby room. He or she will be able to see, hear, and speak with you at all times. You will likely be able to resume your activities right away unless you took a sedative.



## Biopsy

A biopsy is the removal of tissue or fluid samples to test for disease. The samples will be studied under a microscope by a pathologist in order to confirm if there's cancer. A biopsy is recommended for three reasons. First, you may have a biopsy if you will have radiation therapy or chemotherapy before surgery. Second, a biopsy may also be done if your doctor thinks you may have two types of cancer. Third, you may have a biopsy if you won't have surgery. Otherwise, the tumor will be tested for cancer at the time of your surgery.

A core-needle biopsy is recommended. This biopsy removes samples with a needle that is inserted through your skin and into the tumor. Your skin will be numbed beforehand. Because tumors within the abdomen are deep, imaging should be used to guide the needle to the tumor. CT or an ultrasound may be used. Ultrasound makes images with high-frequency sound waves.

A core-needle biopsy is preferred over an open surgical biopsy. An open surgical biopsy requires anesthesia to numb the area and a cut into the skin. It is a minor surgery that removes some but not the entire tumor.

If after the first biopsy it is unclear if the tumor is cancer, a second biopsy should be done. If the tumor is cancer, more testing should be done to learn more about the cancer. Your doctors will want to know the cancer grade. Testing for abnormal genes within the cancer cells may also be done. It is important that the testing be done by a pathologist who has experience with sarcoma.

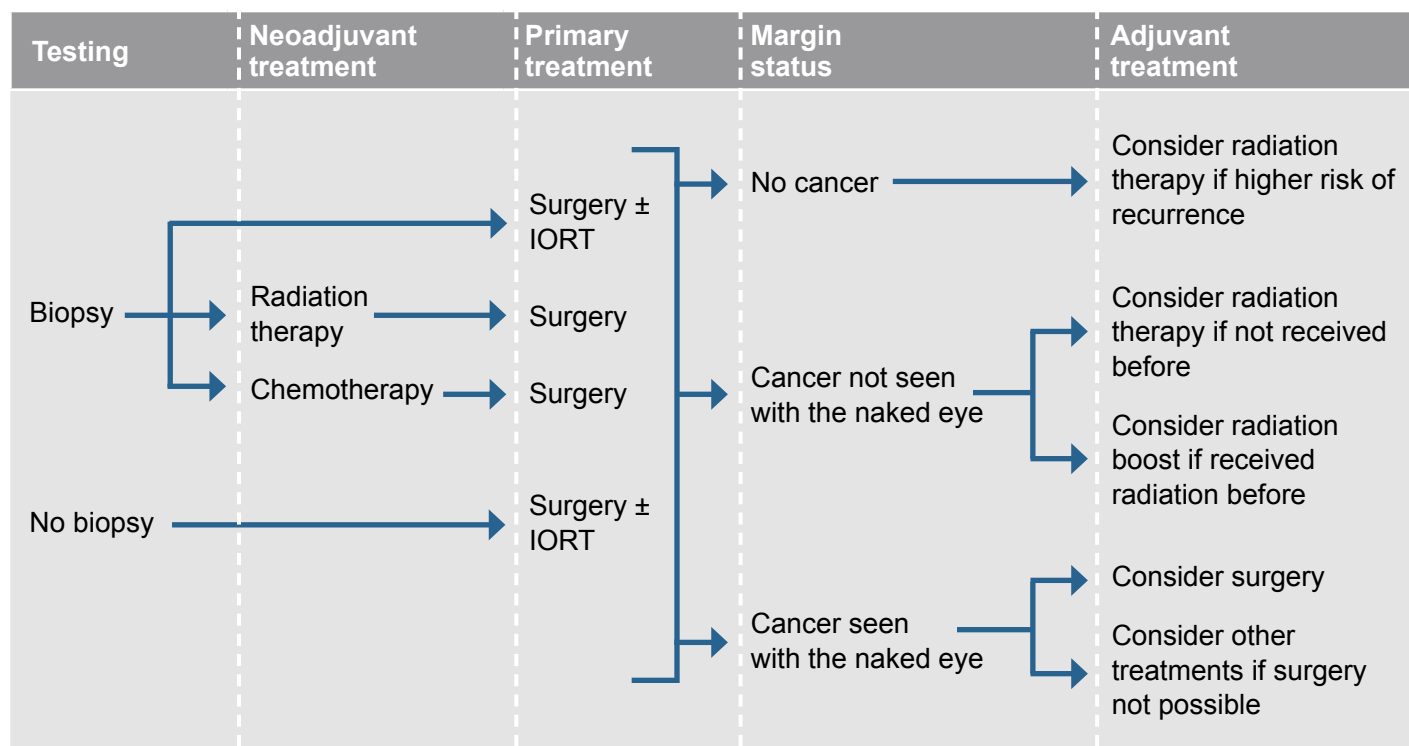
If you may have Li-Fraumeni syndrome, your doctor may want you to have a genetic assessment. A genetics counselor will explain what the goals, benefits, and risks of the assessment are. The counselor may also ask more questions about your

health history as well as the history of your family. You may decide to get tested for Li-Fraumeni syndrome. To be tested, you must provide a sample of body tissue. Using the tissue sample, a pathologist can test your genes for the TP53 mutations that cause Li-Fraumeni syndrome.

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## 3.1 Treatment with surgery

**Chart 3.1.1 Treatment options**



**Chart 3.1.2 Care after treatment**

Type of care	Schedule of care
<ul style="list-style-type: none"> <li>Physical exam and imaging of abdomen and pelvis</li> </ul>	Every 3–6 months for 2–3 years <ul style="list-style-type: none"> <li>◦ If good results, then repeat every 6 months for 2 years</li> <li>◦ If good results, then repeat every year</li> </ul>
<ul style="list-style-type: none"> <li>Consider imaging of chest</li> </ul>	

**Chart 3.1.1** maps the treatment options for sarcomas that can be treated with surgery. Surgery with or without IORT is an option whether you had a biopsy or not. If you had a biopsy, radiation therapy or chemotherapy before surgery is another option. The goal is to remove all the cancer with cancer-free surgical margins.

After surgery, your doctors will assess if you need adjuvant treatment. Adjuvant treatment can 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 cancer isn't found. Only when the cancer is likely to return will your doctors suggest to have radiation therapy.

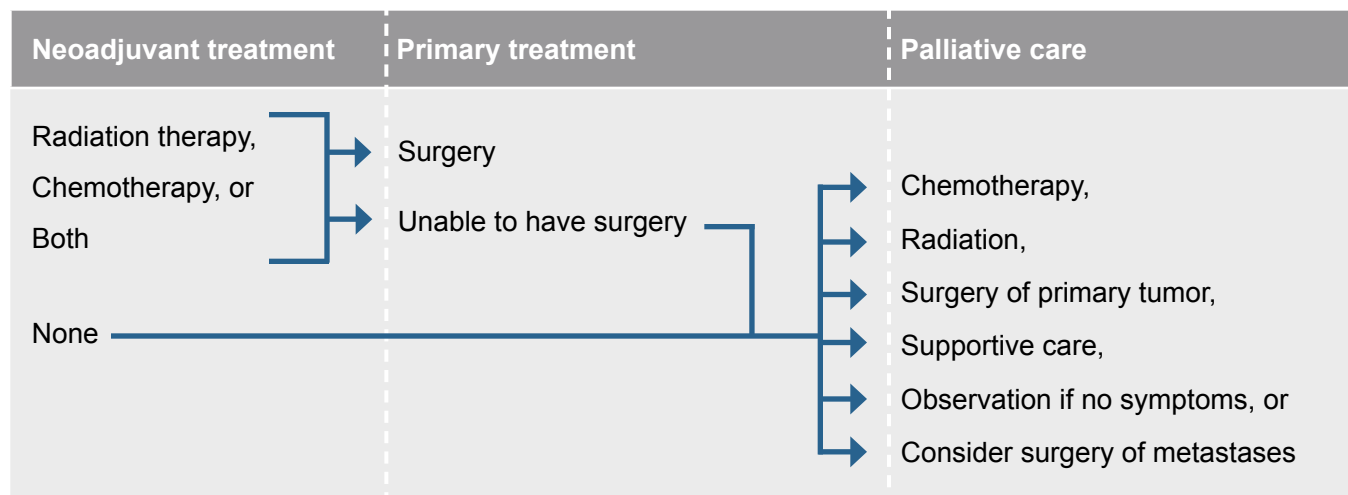
Some margins have cancer cells that aren't seen with the naked eye. Instead, cancer cells are found with a microscope. In this case, you may have radiation therapy to kill any remaining cells. However, if you had radiation therapy before surgery, only a radiation boost may be given.

Many sarcomas are removed with cancer that can be seen at the margin. 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. Read *Treatment without surgery* on page 36 for more information.

**Chart 3.1.2** lists care after cancer treatment has ended. You should have follow-up tests to check if the cancer has returned. Getting follow-up tests can help find cancer early. Cancer is more likely to be cured if found early. 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.

## 3.2 Treatment without surgery

**Chart 3.2 Treatment options**



**Chart 3.2** maps the treatment options for sarcomas that can't be treated with surgery. However, other treatments may shrink the tumor so that surgery can be done. These treatments include radiation therapy, chemotherapy, or both. Radiation may be given if the area was never treated with it before. If after these treatments you are able to have surgery, read *Treatment with surgery* on page 34 for more information.

If you are unable to have surgery, palliative care is an option. Palliative care is also called supportive care. It does not try to cure the cancer but aims to control symptoms.

Chemotherapy, radiation therapy, and surgery may stop or decrease tumor growth. Other supportive care is available depending on your symptoms. If you have no symptoms, observation is an option. Your doctor may also suggest surgery to remove metastatic tumors.

Joining a clinical trial is highly encouraged. You'll likely receive the best management of care. 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.

# Gastrointestinal stromal tumors



# 4 Gastrointestinal stromal tumors

38	Treatment planning
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44	Treatment for other GISTs
46	Treatment after progression
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## Treatment planning

### Must haves

Multidisciplinary team

Medical history and physical exam

Biopsy if

- <2 cm stomach tumor,
- ≥2 cm GI tumor and may have other types of treatment before surgery, or
- ≥2 cm GI tumor and can't have surgery but will have other treatment

Imaging of abdomen and pelvis

- CT (computed tomography) for <2 cm stomach tumors
- CT or MRI (magnetic resonance imaging) for ≥2 cm tumor

KIT and PDGFRA testing for ≥2 cm tumor

### Sometimes useful

Imaging of chest

Endoscopy ± ultrasound

PET (positron emission tomography)

SDH (succinate dehydrogenase) gene testing

This chapter is about GISTs. These tumors may start in cells that trigger muscle movement in the gut wall. Part 4 starts by listing what is needed for treatment planning. Next, treatment options are presented for <2 cm stomach GISTs, all other GISTs, and if GISTs keep growing while taking drug treatment.

### Multidisciplinary team

Treatment of sarcoma takes a team of experts who have experience with this cancer. If you have sarcoma, it is important that all the experts meet before your treatment is started to create the best treatment plan. Your treatment team will also meet while you are going through treatment and afterward to discuss the treatment results and the next steps of care. Your team of experts may include a:

In most cases

- Pathologist—an expert in testing cells and tissue to find 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, and a
- Nurse—an expert trained to care for the sick.

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 live life unaided or with devices,
- Physical therapist—an expert in helping people move better,
- Nutritionist—an expert in healthy foods and drinks, and a
- Genetic counselor—an expert in explaining testing for hereditary diseases.

### Medical history and physical exam

Your medical history includes any health events in your life. It also includes any medications you've taken or are taking. Since some health problems run in families, your doctor will ask about the medical history of your blood relatives.

Doctors often perform a physical exam along with taking a medical history. A physical exam is a review of your body for signs of disease. During this exam, your doctor will listen to your lungs, heart, and gut. Parts of your body will likely be felt to see if organs are of normal size, are soft or hard, or cause pain when touched. Your lymph nodes may feel large if cancer has spread to them.

### Biopsy

GISTs are soft and fragile tumors. Thus, a biopsy called a EUS-FNA (endoscopic ultrasound-guided fine-needle aspiration) is recommended. A biopsy is removal of tissue or fluid samples to test for disease. The samples will be studied under a microscope by a pathologist in order to confirm if there's cancer.

EUS is a better choice of biopsy for GISTs than a biopsy through the skin (percutaneous). With a percutaneous biopsy, there is a chance that the tumor may leak blood (hemorrhage) and spread. A percutaneous biopsy guided to the tumor by imaging may be okay for metastatic tumors.

A biopsy of the tumor should be done for three reasons. First, a biopsy is needed for a stomach tumor smaller than 2 cm. The biopsy can help your doctors know if it is a GIST and decide the best care. Such small tumors may not be an aggressive cancer that needs more treatment.

The second reason for a biopsy is for other GISTs that receive other treatment before surgery. Likewise, a biopsy is recommended when you can't have

surgery but might need other types of treatment. It's important to confirm that the tumor is cancer before starting treatment.

For EUS-FNA, an endoscope will be guided down your esophagus to the tumor. An endoscope is a device with a thin, long tube. At the end of the tube is a light and camera that allows your doctor to see into your body. Endoscopes used for EUS-FNA are fitted with ultrasound, which is a type of imaging test. Ultrasound uses high-frequency sound waves to make pictures. Your doctor will move the ultrasound back and forth to see a 360-degree view of the area on a screen. Once the endoscope is within reach of the tumor, a needle will be used to obtain a sample. You will likely be sedated for EUS-FNA but sometimes anesthesia is used.

If after the first biopsy it is unclear if the tumor is cancer, a second biopsy should be done. If the tumor

is cancer, more testing should be done to learn more about the cancer. Your doctors will want to know the cancer grade. Testing for abnormal genes within the cancer cells may also be done. It is important that the testing be done by a pathologist who has experience with sarcoma.

### Imaging of abdomen and pelvis

If you likely have GIST, you should receive a CT scan of your abdomen and pelvis. CT is a type of imaging test. It takes many x-rays of the same body part from different angles to make detailed images. Your doctor may have you have another imaging test called MRI. MRI uses radio waves and powerful magnets to make images. Imaging can show your doctors how large a tumor is and how close it is to other tissues.

A contrast dye should be used to make the images clearer. The dye will be injected into your vein, mixed with a liquid you drink, or both. The dye may cause you to feel flushed or get hives. Rarely, serious allergic reactions occur. Tell your doctor and the technicians if you have had bad reactions in the past.

Getting an imaging scan is often easy. Before the scan, you may need to stop taking some medicines, stop eating and drinking for a few hours, and remove metal objects from your body. During the scan, you will need to lie face up on a table that moves through the machine. As the machine takes pictures, you may hear buzzing, clicking, or whirring sounds. You will be alone, but a technician will operate the machine in a nearby room. He or she will be able to see, hear, and speak with you at all times. You will likely be able to resume your activities right away unless you took a sedative.

### KIT and PDGFA testing

Many (85 out of 100) GISTs have mutations in the *KIT* gene. KIT is a molecule within a chemical pathway that starts cell growth. It is sometimes called CD117. The mutations in the *KIT* gene cause KIT





to be overactive and the cells to grow fast. Some GISTs that don't have a KIT mutation instead have a mutation in PDGFRA genes. PDGFRA works very much like KIT. GISTs with PDGFRA mutations may have little or no KIT (CD117). Testing of mutations in KIT and PDGFRA genes is strongly recommended. Tissue removed by biopsy or surgical treatment can be used for testing.

### Imaging of chest

Imaging of your chest may be needed. Chest imaging will allow your doctors to assess if the cancer has spread to your lungs. Cancer spread to the lungs is less common for GISTs than other sarcomas. You may receive a plain radiograph or CT of the chest. Chest CTs are often given when images from a radiograph would be unclear due to lung scarring or other health conditions.

### Endoscopy ± ultrasound

Your doctors may want you to have an endoscopy. For this test, an endoscope will be inserted down your mouth and into your digestive tract. Your doctors will be able to assess the size and spread of the tumor. Ultrasound may be used to image tissue that can't be seen with the endoscope.

### PET

PET scans are a type of imaging test. PET scans can help show if a mass is a growing tumor compared to dead or inactive tissue. It can also show changes in chemical activity within a tumor before CT can show changes in shape. However, PET should not be used in place of CT. Many imaging centers have machines that can do both PET and CT.

PET is recommended if you need to start treatment before surgery that would leave you with long-term problems if you didn't have prior treatment. Comparing PET scans taken before and after starting drug treatment may show if treatment is working. Treatment results can be seen within weeks.

To create PET images, a sugar radiotracer is used. If the radiotracer is FDG (fluorodeoxyglucose), you must fast for 4 hours before it is injected into your body. Between 40 to 60 minutes after injection, the radiotracer will emit a small amount of energy. This energy can be detected by the imaging machine.

In the PET images, cancer appears brighter ("hotter"). This is because cancer cells use sugar more quickly than normal cells. Hot spots suggest that cancer is present.

### SDH testing

People who have Carney-Stratakis syndrome are at higher risk for GIST. Mutations in the gene that controls SDH have been linked to people with GIST-related Carney-Stratakis syndrome. If KIT or PDGFRA testing finds no mutations, then your doctors may want to have SDH testing.



## 4.1 Treatment for small stomach GISTs

**Chart 4.1 Treatment options**

Test results	Primary treatment	Follow-up testing
Not high risk	None	Consider endoscopic testing every 6–12 months
High risk	Surgery	Consider CT with contrast of your abdomen and pelvis <ul style="list-style-type: none"> <li>◦ Every 3–6 months for 3–5 years,</li> <li>◦ If normal results, then repeat every year</li> </ul>

**Chart 4.1** maps the treatment options for GISTs of the stomach that are smaller than 2 cm. Many of these tumors are not cancer (benign). However, some will grow fast and will become a problem.

Your doctors will assess the tumor for risk factors that suggest there is a high risk for fast growth. Tumors that aren't high risk don't need treatment. Instead, the tumor can be looked at with an endoscope every 6 to

12 months for changes. Talk to your doctors about the benefits and dangers of such testing.

Surgery to remove the tumor is recommended for those that are at high risk of growing fast. After surgery, you may get CT scans with contrast of your abdomen and pelvis. CT scans every 3 to 6 months for 3 to 5 years is recommended. If test results are normal for 3 to 5 years, CT may be done every year.

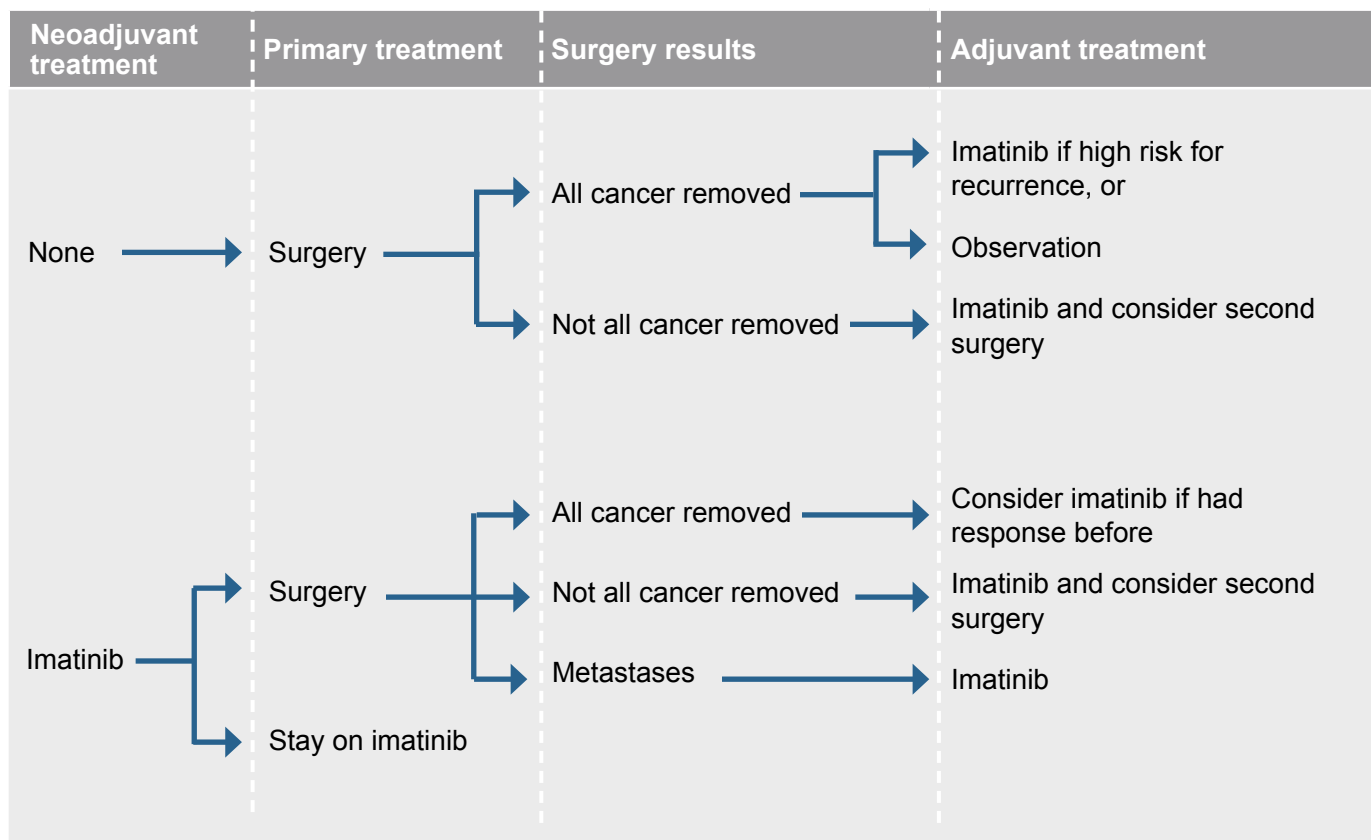


## My notes

[illegible]

## 4.2 Treatment for other GISTs

**Chart 4.2.1 Treatment options**



**Chart 4.2.2 Care after surgery**

Surgery results	Tests	Schedule
All cancer removed	• Medical history and physical exam	Every 3–6 months for 5 years <ul style="list-style-type: none"> <li>◦ If normal results, then repeat every year</li> </ul>
	• CT of abdomen and pelvis	Every 3–6 months for 3–5 years <ul style="list-style-type: none"> <li>◦ If normal results, then repeat every year</li> </ul>
Not all cancer removed	• Medical history, physical exam, and CT of abdomen and pelvis	Every 3–6 months

**Chart 4.2.1** maps 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 as long as it does not leave you with long-term problems afterward. You may receive more treatment after surgery. If all the cancer appears to be removed, you could start imatinib if there's a high risk for the cancer returning. Otherwise, read *Care after surgery* to learn what to do next. If your surgeon was unable to remove all of the cancer, you may start imatinib and consider a second surgery.

## Imatinib first

Imatinib may first be used for GISTs that can be removed by surgery but the surgery would leave you with long-term problems. In this case, imatinib is given to shrink the tumor so that surgery won't cause long-term problems. 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 can show if treatment is working. If the tumor is the same size or smaller, keep taking the same dose of imatinib. Once the tumor is small enough, you may be able to have surgery. After surgery, start imatinib again for adjuvant treatment. If the tumor grows while taking imatinib, surgery may be done for some people. Otherwise, read *Treatment after progression* on page 46 for the next steps of care.

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. Within 3 months of starting imatinib, your doctors will assess if it is working with a CT scan.

If the tumor is the same size or smaller, keep taking imatinib. The tumor may shrink enough for surgery. Your surgeon and medical oncologist should assess if surgery is possible. If you're able to have surgery, imatinib should be restarted afterwards unless your doctor tells you not to. If you're unable to have surgery, stay on imatinib. Read *Treatment after progression* if the tumor grows while on imatinib.

**Chart 4.2.2** lists care for after surgery. You should have follow-up tests. Tests 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. Cancer is more likely to be cured if found 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 every 3 to 6 months for 3 to 5 years. If results are normal during this time, CT is only recommended every year.

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

## 4.3 Treatment after progression

**Chart 4.3.1 First progression**

Cancer growth	Treatment options
Limited	Continue imatinib and consider: <ul style="list-style-type: none"> <li>• Surgery if possible</li> <li>• Ablation, embolization, or chemoembolization</li> <li>• Palliative radiation therapy if cancer spread to your bones</li> </ul> Increase dose of imatinib or change to sunitinib
Widespread	Increase dose of imatinib Change to sunitinib

**Chart 4.3.2 Second progression**

Treatment options
<ul style="list-style-type: none"> <li>• Regorafenib</li> <li>• Clinical trial</li> <li>• Alternate treatments (little research done)</li> <li>• Supportive care</li> </ul>

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

**Chart 4.3.1** 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.

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 may be used if CT or MRI results are unclear.

Joining a clinical trial is highly encouraged. You'll likely receive the best management of care. Ask your treatment team if there are clinical trials you can join.

**Chart 4.3.2** lists treatment options if the cancer keeps growing while taking imatinib or sunitinib. There are four treatment options. Taking regorafenib is strongly supported. You may also join a clinical trial or start alternate treatment. However, less research has been done to know if the alternate treatments will be helpful. 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. The tumor will grow more slowly on some treatment than on none.

- GISTs may start in cells that trigger muscle movement in the gut wall.
- Treatment planning is an important first step of care.
- 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 may be first 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.

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# Desmoid tumors



# 5 Desmoid tumors

50	Treatment planning
53	Treatment with surgery
55	Treatment without surgery
56	Review



Desmoid tumors aren't cancer but act somewhat like cancer. These tumors will grow into nearby tissue but won't spread to distant sites. This chapter starts by listing what is needed for treatment planning. Treatment options are then presented for when surgery can and can't be done. What you should do after treatment concludes the chapter.

## Treatment planning

### Must haves

- Multidisciplinary team
- Medical history and physical exam
- Imaging of tumor

### Sometimes useful

- Biopsy



## Multidisciplinary team

Treatment of desmoid tumors takes a team of experts who have experience with this disease. If you have a desmoid tumor, it is important that all the experts meet before your treatment is started to create the best treatment plan. Your treatment team will also meet while you are going through treatment and afterward to discuss the treatment results and the next steps of care. Your team of experts may include a:

In most cases

- Pathologist—an expert in testing cells and tissue to find 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, and a
- Nurse—an expert trained to care for the sick.

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 live life unaided or with devices,
- Physical therapist—an expert in helping people move better,
- Nutritionist—an expert in healthy foods and drinks, and a
- Genetic counselor—an expert in explaining testing for hereditary diseases.

## Medical history and physical exam

Your medical history includes any health events in your life. It also includes any medications you've taken or are taking. Since some health problems run in families, your doctor will ask about the medical history of your blood relatives.

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 (familial adenomatous polyposis) 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.

Doctors often perform a physical exam along with taking a medical history. A physical exam is a review of your body for signs of disease. During this exam, your doctor will listen to your lungs, heart, and gut. Parts of your body will likely be felt to see if organs are of normal size, are soft or hard, or cause pain when touched.

## Imaging of tumor

Imaging tests make pictures (images) of the insides of your body. Imaging of the tumor is recommended. You may receive CT or MRI. CT takes many x-rays of the same body part from different angles to make detailed images. MRI uses radio waves and powerful magnets to make images. These tests can show your doctors how large a tumor is and how close it is to other tissues.

A contrast dye may be used to make the images clearer. The dye will be injected into your vein, mixed with a liquid you drink, or both. The dye may cause you to feel flushed or get hives. Rarely, serious allergic reactions occur. Tell your doctor and the technicians if you have had bad reactions in the past.

Getting an imaging scan is often easy. Before the scan, you may need to stop taking some medicines, stop eating and drinking for a few hours, and remove metal objects from your body. During the scan, you will need to lie face up on a table that moves through the machine.

As the machine takes pictures, you may hear buzzing, clicking, or whirring sounds. You will be alone, but a technician will operate the machine in a nearby room. He or she will be able to see, hear, and speak with you at all times. You will likely be able to resume your activities right away unless you took a sedative.

### Biopsy

A biopsy is the removal of tissue or fluid samples to test for disease. After the biopsy, a pathologist will examine the samples with a microscope to assess if there's cancer. A biopsy is recommended unless you will have surgery that fully removes the tumor.

There are multiple ways that a biopsy can be done. A needle can be used to remove samples. The two types of needle biopsies are a core-needle biopsy and fine-needle aspiration. The core-needle biopsy obtains a larger sample for testing. A fine-needle aspiration removes a smaller piece of tissue and doesn't require any cutting into the skin (incision).

Besides needle biopsies, an incisional biopsy can be used. An incisional biopsy requires anesthesia to numb the area and cutting into the skin. It is a minor surgery that removes some but not the entire tumor.

If after the first biopsy it is unclear if the tumor is cancer, a second biopsy should be done. The second biopsy should be done using imaging to guide the needle into the tumor. If you have cancer and not a desmoid tumor, more testing should be done to learn more about the cancer. Your doctors will want to know the cancer grade. Testing for abnormal genes within the cancer cells may also be done. It is important that the testing be done by a pathologist who has experience with sarcoma.

## 5.1 Treatment with surgery

**Chart 5.1.1 Treatment options**

Primary treatment	Treatment results	Adjuvant treatment
Observation if small tumor		
Options for all tumors:		
Surgery ± radiation therapy ± drug therapy	Tumor fully gone	Observation, or Consider radiation if large tumor
Radiation therapy	Some tumor remains but can't be seen	Consider second surgery, Radiation therapy if none prior, or Observation
Drug therapy	Some tumor remains and can be seen	Radiation therapy, Drug therapy, Surgery if all else fails Observation

**Chart 5.1.1** maps 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 and the possible outcomes of treatment. Options include surgery with or without radiation therapy, chemotherapy, or both. Other options are radiation therapy or drug therapy without other treatments.

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 a second surgery, radiation therapy, or observation.

Tumors that can still be seen after treatment have four options for treatment. Radiation and drug therapy are two options. If neither works, surgery to remove the visible tumor may be done. Lastly, observation can be done, and when the tumor grows you can start treatment.

**Chart 5.1.2 Care after treatment**

Type of care	Schedule of care
• Rehabilitation	If needed
• Report any new or worse symptoms to your doctors	
• Medical history, physical exam, and imaging	Every 3–6 months for 2–3 years ▫ If good results, then repeat every year

**Chart 5.1.2** lists care after treatment 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. If results are normal during this time, these tests can be done every year.

## 5.2 Treatment without surgery

**Chart 5.2.1 Treatment options**

- Radiation therapy
- Drug therapy
- Surgery if all else fails
- Observation

**Chart 5.2.2 Care after treatment**

Type of care	Schedule of care
• Rehabilitation	If needed
• Report any new or worse symptoms to your doctors	
• Medical history, physical exam, and imaging	Every 3–6 months for 2–3 years <ul style="list-style-type: none"> <li>▫ If good results, then repeat every year</li> </ul>

**Chart 5.2.1** lists recommended options for tumors that can't be fully removed with surgery. There are four treatment options. Radiation and drug therapy are two options. If neither works, surgery to remove the visible tumor may be done. Last, observation can be done, and when the tumor grows you can start treatment.

**Chart 5.2.2** lists care after treatment 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. If results are normal during this time, these tests can be done every year.

- Desmoid tumors aren't cancer but act somewhat like cancer.
- Treatment planning is an important first step of care.

- Some desmoid tumors can be treated with surgery while others cannot.
- Receiving care after treatment is important to find any new tumors early.

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# Making treatment decisions



# 6 Making treatment decisions

58	It's your choice
59	Questions to ask your doctors
63	Weighing your options
64	Websites   Review



Parts 1 through 5 described the cancer and gave test and treatment options recommended by NCCN experts. These options are based on science and agreement among NCCN experts. Part 6 aims to help you make decisions that are in line with your beliefs, wishes, and values.

## 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'.

Your doctors will give you the information you need to make an informed choice. However, letting others decide which option is best may make you feel more at ease. But, who do you want to make the decisions? You may rely on your doctors alone to make the right decisions. You can also have loved ones help. They can gather information, speak on your behalf, and share decision-making with your doctors. Even if others decide the best option, you still have to agree to have this treatment by signing a consent form.

On the other hand, you may want to take the lead or share in decision-making. Many patients do. In shared decision-making, you and your doctors share information, weigh the options, and agree on a treatment plan. Your doctors know the science but you know your concerns and goals. By working together, you are likely to get higher-quality care and be more satisfied. You'll likely get the treatment you want, at the place you want, and by the doctors you want.



## Questions to ask your doctors

You will likely meet with experts from different fields of medicine. Strive to have helpful talks with each person. Prepare questions before your visit and ask questions if the person isn't clear. You can also record your talks and get copies of your medical

records. It may be helpful to have your spouse, partner, family member, or a friend with you at these visits. They can help to ask questions and remember what was said. Suggested questions to ask include:

## What's my diagnosis and prognosis?

It's important to know that there are different types of cancer. Cancer can greatly differ even when people have a tumor in the same organ. Based on your test results, your doctors can tell you which type of cancer you have. They can also give a prognosis. A prognosis is a prediction of the pattern and outcome of a disease. Knowing the prognosis may affect what you decide about treatment.

1. Where did the cancer start? In what type of cell?
2. Is this cancer common?
3. What is the cancer stage? Does this stage mean the cancer has spread far?
4. What is the grade of the cancer? Does this grade mean the cancer will grow and spread fast?
5. What other test results are important to know?
6. How often are these tests wrong?
7. Would you give me a copy of the pathology report and other test results?
8. Can the cancer be cured? If not, how well can treatment stop the cancer from growing?

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## What are my options?

There is no single treatment practice that is best for all patients. There is often more than one treatment option along with clinical trial options. Your doctor will review your test results and recommend treatment options.

1. What will happen if I do nothing?
2. Can I just carefully monitor the cancer?
3. Should I consider a clinical trial?
4. Do you consult NCCN recommendations when considering options?
5. Are you suggesting options other than what NCCN recommends? If yes, why?
6. How do my age, health, and other factors affect my options?
7. Which option is proven to work best?
8. Which options lack scientific proof?
9. What are the benefits of each option? Does any option offer a cure? Are my chances any better with one option than another? Which option spares the most healthy tissue? Is any option less invasive? Less time-consuming? Less expensive?
10. What are the risks of each option? What are possible complications? What are the rare and common side effects? Short-lived and long-lasting side effects? Serious or mild side effects? Other risks?

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## What does each option require of me?

Many patients consider how each option will practically affect their lives. This information may be important because you have family, jobs, and other duties to take care of. You may also be concerned about getting the help you need. If you have more than one option, choosing the option that is the least taxing may be important to you.

1. Will I have to go to the hospital or elsewhere? How many times? How long is each visit?
2. How do I prepare for treatment?
3. Should I bring someone with me when I get treated?
4. Will the treatment hurt?
5. How much will the treatment cost me?
6. Is home care after treatment needed? If yes, what type?
7. How soon will I be able to manage my own health?
8. When will I be able to return to my normal activities?

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More and more research is finding that patients treated by more experienced doctors have better results. It is important to learn if a doctor is an expert in the cancer treatment he or she is offering.

1. Are you board certified? If yes, in what area?
2. How many patients 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?

This image shows a single sheet of white paper with horizontal ruling lines. The lines are evenly spaced and run across the width of the page. There are no margins, text, or other markings on the paper.

## Weighing your options

Deciding which option is best can be hard. Doctors from different fields of medicine may differ 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.

### 2<sup>nd</sup> opinion

The time around a cancer diagnosis is very stressful. People with cancer often want to get treated as soon as possible. They want to make their cancer go away before it spreads farther. While cancer can't be ignored, there is time to think about and choose which option is best for you.

You may wish to have another doctor review your test results and suggest a treatment plan. This is called getting a 2<sup>nd</sup> opinion. You may completely trust your doctor, but a 2<sup>nd</sup> opinion on which option is best can help.

Copies of the pathology report, a DVD of the imaging tests, and other test results need to be sent to the doctor giving the 2<sup>nd</sup> opinion. Some people feel uneasy asking for copies from their doctors. However, a 2<sup>nd</sup> opinion is a normal part of cancer care.

When doctors have cancer, most will talk with more than one doctor before choosing their treatment. What's more, some health plans require a 2<sup>nd</sup> opinion. If your health plan doesn't cover the cost of a 2<sup>nd</sup> opinion, you have the choice of paying for it yourself.

If the two opinions are the same, you may feel more at peace about the treatment you accept to have. If the two opinions differ, think about getting a 3<sup>rd</sup> opinion. A 3<sup>rd</sup> opinion may help you decide between

your options. Choosing your cancer treatment is a very important decision. It can affect your length and quality of life.

### Support groups

Besides talking to health experts, it may help to talk to patients who have walked in your shoes. Support groups often consist of people at different stages of treatment. Some may be in the process of deciding while others may be finished with treatment. At support group meetings, you can ask questions and hear about the experiences of other patients.

### Compare benefits and downsides

Every option has benefits and downsides. Consider these when deciding which option is best for you. Talking to others can help identify benefits and downsides you haven't thought of. Scoring each factor from 0 to 10 can also help since some factors may be more important to you than others.



## Websites

### American Cancer Society

[www.cancer.org/cancer/sarcoma-adultsofttissuecancer](http://www.cancer.org/cancer/sarcoma-adultsofttissuecancer)

### National Cancer Institute

[www.cancer.gov/cancertopics/pdq/treatment/adult-soft-tissue-sarcoma/Patient](http://www.cancer.gov/cancertopics/pdq/treatment/adult-soft-tissue-sarcoma/Patient)

### Sarcoma Alliance

[sarcomaalliance.org](http://sarcomaalliance.org)

### Sarcoma Alliance for Research through Collaboration

[www.sarctrials.org](http://www.sarctrials.org)

### Sarcoma Foundation of America

[www.curesarcoma.org](http://www.curesarcoma.org)

## Review

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

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# Glossary

Dictionary

Acronyms

Types of sarcoma

# Dictionary

**abdomen**

The belly area between the chest and pelvis.

**ablation**

Treatment that destroys very small tumors.

**adjuvant treatment**

Treatment that is given after the main treatment used to cure the cancer.

**allergic reaction**

Symptoms that are caused when the body is trying to rid itself of outside agents.

**angiogram**

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

**biopsy**

Removal of small amounts of tissue or fluid to be tested for disease.

**brachytherapy**

Treatment with radioactive objects placed near or in a tumor. Also called internal radiation.

**cancer grade**

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

**cancer stage**

A rating of the growth and spread of cancer.

**Carney-Stratakis syndrome**

A health condition that is passed down from parents and marked by gastrointestinal stromal tumors.

**catheter**

A flexible tube inserted in the body to give treatment or drain fluid from the body.

**chemoembolization**

Treatment that cuts off blood supply to tumors with beads coated with chemotherapy.

**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**

Research on a test or treatment to assess its safety or how well it works.

**computed tomography (CT)**

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

**contrast**

A dye that is put into the body to make clearer pictures during imaging tests.

**core-needle biopsy**

Removal of a large tissue sample with a thick, hollow needle to test for disease.

**deoxyribonucleic acid (DNA)**

A chain of chemicals inside cells that contains coded instructions for making and controlling cells.

**desmoid tumor**

A mass of fibrous cells that can grow into nearby tissue but can't spread to distant sites.

**embolization**

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

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

Removal of fluid with a needle that is guided with an imaging test to the tumor.

**external beam radiation therapy (EBRT)**

Treatment with radiation received from a machine outside the body.

**familial adenomatous polyposis (FAP)**

A health condition that is passed down from parents and increases the chance of getting sarcoma.

**fascia**

A deep layer of soft tissue.



**fine-needle aspiration (FNA)**

Use of a thin needle to remove fluid or tissue from the body to test for disease.

**Gardner's syndrome**

A health condition that is passed down from parents and increases the chance of getting sarcoma.

**gastroenterologist**

A doctor who's an expert in digestive diseases.

**gene**

A set of coded instructions in cells needed to make new cells and control how cells behave.

**general anesthesia**

A controlled loss of wakefulness from drugs.

**genetic assessment**

Testing for diseases that are caused by abnormal information in cells that is passed down from parents.

**hereditary**

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

**hives**

Itchy, swollen, and red skin caused by the body trying to rid itself of an outside agent.

**imaging**

A test that makes pictures of the insides of the body.

**immune system**

The body's natural defense against disease.

**immunotherapy**

Treatment that uses the body's natural defense against disease.

**intensity-modulated radiation therapy (IMRT)**

Treatment with radiation that uses small beams of different strengths based on the thickness of the tissue.

**interoperative radiation therapy (IORT)**

Radiation therapy given during surgery.

**isolated limb chemotherapy**

A method of giving drugs through a needle directly into a leg or arm.

**KIT**

A molecule within a chemical pathway that starts cell growth.

**Li-Fraumeni syndrome**

A health condition that is passed down from parents and increases the chance of getting sarcoma.

**lymph node**

A small group of disease-fighting cells.

**magnetic resonance imaging (MRI)**

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

**medical history**

All health events and medications taken to date.

**medical oncologist**

A doctor who's an expert in cancer drugs.

**metastasectomy**

Surgery to remove tumors that formed far from the first site of cancer.

**metastasis**

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

**mutation**

An abnormal change in a cell's coded instructions for making and controlling cells.

**neoadjuvant treatment**

Treatment that is given before the main treatment used to cure a disease.

**nutritionist**

An expert in healthy foods and drinks.

**observation**

A period of testing for cancer growth.

**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**

Treatment for symptoms of a disease. Also sometimes called supportive care.

**pathologist**

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

**PDGFRA**

A molecule within a chemical pathway that starts cell growth.

**pelvis**

The area of the body between the hipbones.

**physical exam**

A review of the body by a health expert for signs of disease.

**physical therapist**

An expert in helping people move better.

**plain radiograph**

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

**plastic surgeon**

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

**positron emission tomography (PET)**

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

**primary treatment**

The main treatment used to rid the body of cancer.

**primary tumor**

The first mass of cancer cells in the body.

**prognosis**

The expected pattern and outcome of a disease based on tests.

**progression**

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

**radiation oncologist**

A doctor who's an expert in radiation treatment.

**radiation therapy**

The use of radiation to treat cancer.

**radiologist**

A doctor who's an expert in imaging tests.

**retroperitoneum**

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

**sarcoma**

Cancer that starts in bones or soft tissue of the body.

**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**

Cancer that starts in tissue that supports, connects, and surrounds parts of your body.

**stereotactic body radiation therapy (SBRT)**

Treatment with radiation that is delivered with precise, high-dose beams.

**succinate dehydrogenase (SDH)**

A protein within cells.

**supportive care**

Treatment for symptoms of a disease. 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**

Treatment with drugs that target a specific or unique feature of cancer cells.

**thoracic surgeon**

A doctor who's an expert in operations within the chest.

**Three-dimensional conformal radiation therapy (3D-CRT)**

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

**TP53**

An abnormal change in cells that causes Li-Fraumeni syndrome.

**ultrasound**

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

# Acronyms

## 3D-CRT

three-dimensional conformal radiation therapy

## CT

computed tomography

## DNA

deoxyribonucleic acid

## EBRT

external beam radiation therapy

## EUS

endoscopic ultrasound-guided fine-needle aspiration

## FAP

familial adenomatous polyposis

## GIST

gastrointestinal stromal tumors

## 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

## PET

positron emission tomography

## SBRT

stereotactic body radiation therapy

## SDH

succinate dehydrogenase

## NCCN Abbreviations and Acronyms

### NCCN®

National Comprehensive Cancer Network®

### NCCN Patient Guidelines®

NCCN Guidelines for Patients®

### NCCN Guidelines®

NCCN Clinical Practice Guidelines in Oncology®

# Types of sarcoma

Alveolar soft-part sarcoma

Angiosarcoma

Hemangiosarcomas

Lymphangiosarcomas

Clear cell sarcoma

Dermatofibrosarcoma protuberans

Desmoplastic small round cell tumor

Epithelioid sarcoma

Fibrosarcoma

Gastrointestinal stromal tumors

Hemangiopericytoma

Kaposi's sarcoma

Leiomyosarcoma

Liposarcoma

Malignant fibrous histiocytoma

Malignant hemangiopericytoma

Malignant peripheral nerve sheath tumor

Malignant schwannomas

Myxofibrosarcoma

Neurofibrosarcomas

Neurogenic sarcomas

Pleomorphic sarcoma, not otherwise specified

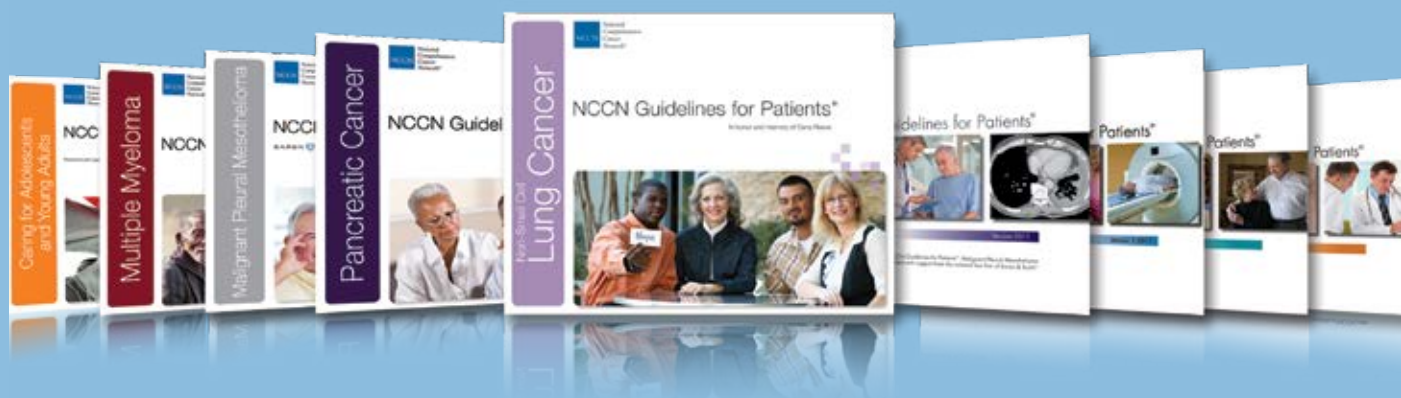
Rhabdomyosarcoma

Synovial sarcoma

# NCCN Guidelines for Patients®

The same authoritative sources referenced by physicians and other health care professionals are available for patients:

- Caring for Adolescents and Young Adults
- Chronic Myelogenous Leukemia
- Colon Cancer
- Esophageal Cancer
- Lung Cancer Screening
- Malignant Pleural Mesothelioma
- Melanoma
- Multiple Myeloma
- Non-Small Cell Lung Cancer
- Ovarian Cancer
- Pancreatic Cancer
- Prostate Cancer
- Stage 0 Breast Cancer
- Stages I and II Breast Cancer
- Stage III Breast Cancer
- Stage IV Breast Cancer



Available at [NCCN.org/patients](http://NCCN.org/patients)

To request a printed copy, e-mail: [patientguidelines@nccn.org](mailto:patientguidelines@nccn.org)



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800.999.5465  
[nebraskamed.com/cancer](http://nebraskamed.com/cancer)

City of Hope Comprehensive Cancer Center  
Los Angeles, California  
800.826.4673  
[cityofhope.org](http://cityofhope.org)

Dana-Farber/Brigham and Women's Cancer Center  
Massachusetts General Hospital Cancer Center  
Boston, Massachusetts  
877.332.4294  
[dfbwc.org](http://dfbwc.org)  
[massgeneral.org/cancer](http://massgeneral.org/cancer)

Duke Cancer Institute  
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[dukecancerinstitute.org](http://dukecancerinstitute.org)

Fox Chase Cancer Center  
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[foxchase.org](http://foxchase.org)

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877.585.0303  
[huntsmancc.org](http://huntsmancc.org)

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Seattle Cancer Care Alliance  
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206.288.7222 • [seattlecca.org](http://seattlecca.org)  
206.667.5000 • [fhcrc.org](http://fhcrc.org)

The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins  
Baltimore, Maryland  
410.955.8964  
[hopkinskimmelcancercenter.org](http://hopkinskimmelcancercenter.org)

Robert H. Lurie Comprehensive Cancer Center of Northwestern University  
Chicago, Illinois  
866.587.4322  
[cancer.northwestern.edu](http://cancer.northwestern.edu)

Mayo Clinic Cancer Center  
Phoenix/Scottsdale, Arizona  
Jacksonville, Florida  
Rochester, Minnesota  
800.446.2279 • Arizona  
904.953.0853 • Florida  
507.538.3270 • Minnesota  
[mayoclinic.org/departments-centers/mayo-clinic-cancer-center](http://mayoclinic.org/departments-centers/mayo-clinic-cancer-center)

Memorial Sloan Kettering Cancer Center  
New York, New York  
800.525.2225  
[mskcc.org](http://mskcc.org)

Moffitt Cancer Center  
Tampa, Florida  
800.456.3434  
[moffitt.org](http://moffitt.org)

The Ohio State University Comprehensive Cancer Center - James Cancer Hospital and Solove Research Institute  
Columbus, Ohio  
800.293.5066  
[cancer.osu.edu](http://cancer.osu.edu)

Roswell Park Cancer Institute  
Buffalo, New York  
877.275.7724  
[roswellpark.org](http://roswellpark.org)

Siteman Cancer Center at Barnes-Jewish Hospital and Washington University School of Medicine  
St. Louis, Missouri  
800.600.3606  
[siteman.wustl.edu](http://siteman.wustl.edu)

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901.683.0055 • [westclinic.com](http://westclinic.com)

Stanford Cancer Institute  
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877.668.7535  
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Birmingham, Alabama  
800.822.0933  
[www3.ccc.uab.edu](http://www3.ccc.uab.edu)

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[cancer.ucsd.edu](http://cancer.ucsd.edu)

UCSF Helen Diller Family Comprehensive Cancer Center  
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800.689.8273  
[cancer.ucsf.edu](http://cancer.ucsf.edu)

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University of Michigan Comprehensive Cancer Center  
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800.865.1125  
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Vanderbilt-Ingram Cancer Center  
Nashville, Tennessee  
800.811.8480  
[vicc.org](http://vicc.org)

Yale Cancer Center/  
Smilow Cancer Hospital  
New Haven, Connecticut  
855.4.SMILOW  
[yalecancercenter.org](http://yalecancercenter.org)

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# Soft Tissue Sarcoma

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