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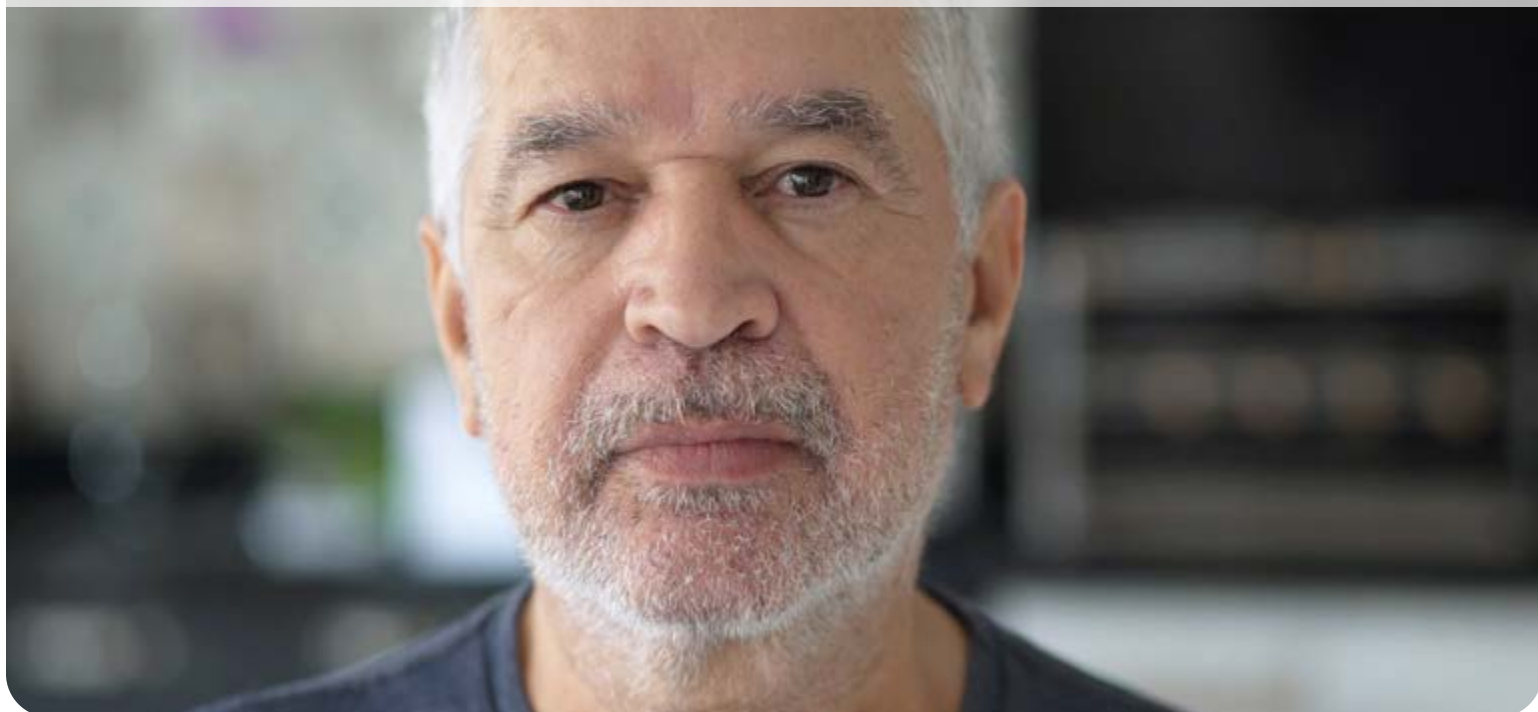
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# Soft Tissue Sarcoma

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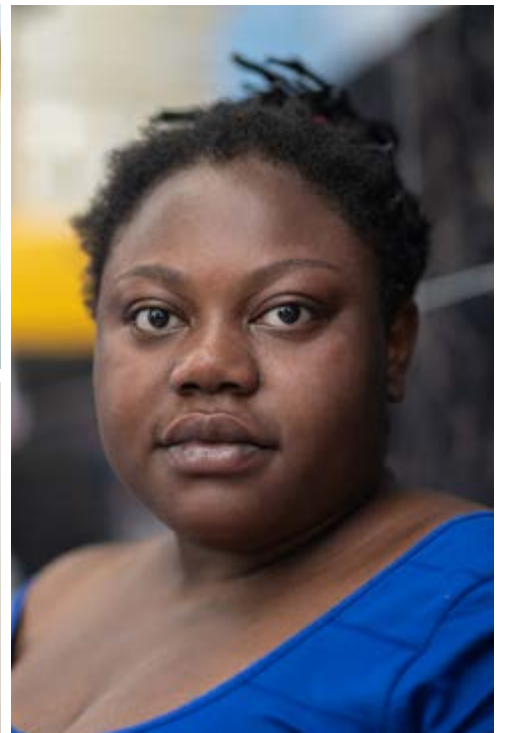
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## Contents

6	Sarcoma basics
13	Testing for soft tissue sarcoma
21	Treatment options
30	Sarcomas in the limbs, outer torso, head, or neck
50	Sarcomas inside the torso
58	Gastrointestinal stromal tumors
71	Desmoid tumors
77	Rhabdomyosarcoma
84	Making treatment decisions
93	Sarcoma mutations
100	Words to know
104	NCCN Contributors
105	NCCN Cancer Centers
106	Index

# 1

## Sarcoma basics

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7	Sarcoma
8	Soft tissue sarcoma
12	Risk factors
12	Review





Soft tissue sarcoma is a rare type of cancer that begins in the soft tissue. Soft tissue is found everywhere in the body. It includes muscle, fat, nerves, tendons, blood and lymph vessels, and tissue around joints. There are over 50 types of soft tissue sarcoma.

## Sarcoma

Sarcomas are a large but rare group of cancers that start in the connective tissue of the body. Connective tissue includes bones, cartilage, muscles, tendons, veins, arteries, and nerves. These tissues support, protect, and give structure to other tissues and organs in the

body. Connective tissue not only holds things together, but is also stores fat, helps move nutrients, and repairs damaged tissue.

Sarcomas are divided into 2 main types:

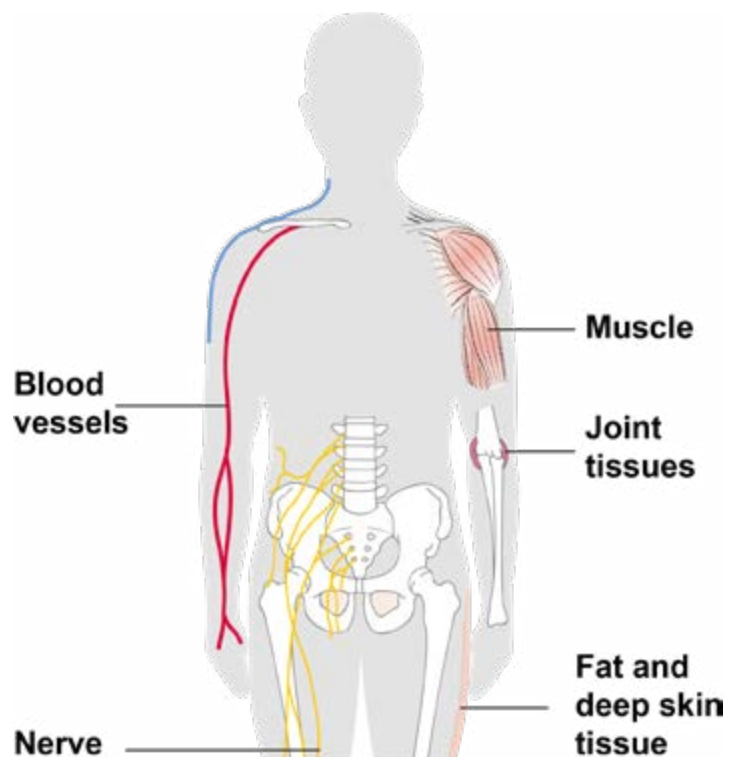
- Soft tissue
- Bone

Soft tissue includes fat, muscles, nerves, tendons, blood and lymph vessels, and other supportive tissues of the body. Soft tissue sarcomas are more common than bone sarcomas. Bone sarcomas, such as osteosarcomas, start in the cells that form bone. Soft tissue sarcomas start in the cells of different types of soft tissue. Soft tissue sarcoma is the focus of this book.

### Soft tissue

**Soft tissue is found everywhere in the body. It includes fat, muscles, nerves, blood and lymph vessels, and tissues surrounding joints.**

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## Soft tissue sarcoma

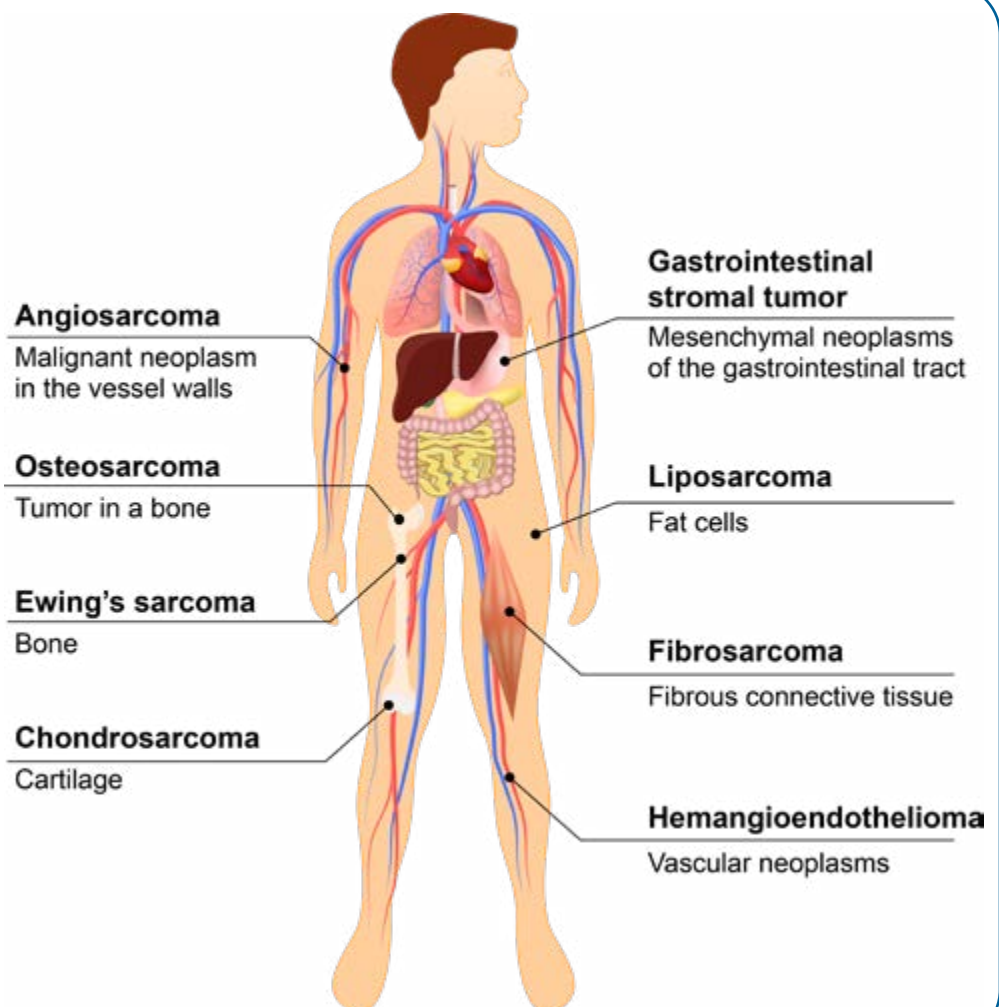
Cancer is a disease that starts in the cells of your body. Soft tissue sarcoma starts in the mesenchymal cells that develop into soft tissue.

There are over 50 types of soft tissue sarcoma that can be found anywhere in the body. This makes it a challenge to diagnose and treat. Since soft tissue sarcomas are very rare, many doctors are not experts in diagnosing or treating these tumors. Therefore, it is important to find a treatment center or hospital that has both experts and experience in your type of soft tissue sarcoma.

Treatment is based on the type and location of the soft tissue sarcoma. The most common site for a sarcoma is in the extremities, known as limbs (arms and legs). The second most common site is in the abdomen (belly), such as the stomach and intestines. Other areas include the space in front of your lower spine called the retroperitoneum, the trunk, and the head or neck. The trunk is also called the torso. It is the main part of the body not including the head, neck, or limbs.

### Types of sarcoma

There are many types of sarcoma. This image shows only some types of sarcoma.



## Histology

The study of the anatomy (structure) of cells, tissues, and organs under a microscope is called histology or histopathology. The cells of each type of sarcoma look different under a microscope. An accurate histology is an important part of a diagnosis and treatment plan.

Some types of soft tissue sarcoma are:

- Undifferentiated pleomorphic sarcoma – the cells don't look like the soft tissue in which they are found
- Gastrointestinal stromal tumors (GISTs) - form in the digestive tract
- Liposarcoma – forms in fat
- Leiomyosarcoma – forms in smooth muscle
- Malignant peripheral nerve sheath tumors – form in nerves

Since there are many types of soft tissue sarcoma that can appear anywhere in the body, diagnosis and treatment are based on the type and location. For example, liposarcomas and undifferentiated pleomorphic sarcomas are most common in the arms and legs. Leiomyosarcomas are most often found in the abdomen.

Soft tissue sarcomas in this book are divided into:

- Limbs, outer torso, head, or neck
- Retroperitoneal and intra-abdominal (inside the abdomen and inner torso)
- Gastrointestinal stromal tumors (GISTs)
- Desmoid tumors (aggressive fibromatosis)
- Rhabdomyosarcoma (RMS)

Some soft tissue sarcomas are so rare that there aren't specific treatment recommendations. Therefore, it is important to be diagnosed and treated by a team of doctors from different fields of medicine who have knowledge and experience in your specific type of soft tissue sarcoma. [See Guide 1 and 2.](#)

Those with soft tissue sarcoma should be treated by a team of doctors from different fields of medicine who have knowledge and experience with your type of soft tissue sarcoma.

## Guide 1

## Types of soft tissue sarcoma as listed by the 2013 World Health Organization

<b>Adipocytic tumors</b>	<ul style="list-style-type: none"> <li>• Atypical lipomatous tumor/well-differentiated liposarcoma</li> <li>• Liposarcoma, NOS</li> </ul>	<ul style="list-style-type: none"> <li>• Dedifferentiated liposarcoma</li> <li>• Myxoid/round cell liposarcoma</li> <li>• Pleomorphic liposarcoma</li> </ul>
<b>Fibroblastic/ myofibroblastic tumors</b>	<ul style="list-style-type: none"> <li>• Dermatofibrosarcoma protuberans</li> <li>• Fibrosarcomatous dermatofibrosarcoma protuberans</li> <li>• Pigmented dermatofibrosarcoma protuberans</li> <li>• Solitary fibrous tumor, malignant</li> <li>• Inflammatory myofibroblastic tumor</li> </ul>	<ul style="list-style-type: none"> <li>• Low-grade myofibroblastic sarcoma</li> <li>• Adult fibrosarcoma</li> <li>• Myxofibrosarcoma (formerly myxoid malignant fibrous histiocytoma [myxoid MFH])</li> <li>• Low-grade fibromyxoid sarcoma</li> <li>• Sclerosing epithelioid fibrosarcoma</li> </ul>
<b>So-called fibrohistiocytic tumors</b>	<ul style="list-style-type: none"> <li>• Giant cell tumor of soft tissues</li> </ul>	
<b>Smooth muscle tumors</b>	<ul style="list-style-type: none"> <li>• Leiomyosarcoma (excluding skin)</li> </ul>	
<b>Pericytic (perivascular) tumors</b>	<ul style="list-style-type: none"> <li>• Malignant glomus tumor</li> </ul>	
<b>Skeletal muscle tumors</b>	<ul style="list-style-type: none"> <li>• Embryonal rhabdomyosarcoma (including botryoid, anaplastic)</li> <li>• Alveolar rhabdomyosarcoma (including solid, anaplastic)</li> <li>• Pleomorphic rhabdomyosarcoma</li> <li>• Spindle cell/sclerosing rhabdomyosarcoma</li> </ul>	
<b>Vascular tumors</b>	<ul style="list-style-type: none"> <li>• Retiform hemangioendothelioma</li> <li>• Pseudomyogenic (epithelioid sarcoma-like) hemangioendothelioma</li> <li>• Epithelioid hemangioendothelioma</li> <li>• Angiosarcoma of soft tissue</li> </ul>	

## Guide 2

## Types of soft tissue sarcoma (continued)

<b>Chondro-osseous tumors</b>	<ul style="list-style-type: none"> <li>• Extraskeletal osteosarcoma</li> </ul>
<b>Gastrointestinal stromal tumors</b>	<ul style="list-style-type: none"> <li>• Gastrointestinal stromal tumor, malignant</li> </ul>
<b>Nerve sheath tumors</b>	<ul style="list-style-type: none"> <li>• Malignant peripheral nerve sheath tumor</li> <li>• Epithelioid malignant peripheral nerve sheath tumor</li> <li>• Malignant triton tumor</li> <li>• Malignant granular cell tumor</li> </ul>
<b>Tumors of uncertain differentiation</b>	<ul style="list-style-type: none"> <li>• Ossifying fibromyxoid tumor, malignant</li> <li>• Stromal sarcoma, NOS</li> <li>• Myoepithelial carcinoma</li> <li>• Phosphaturic mesenchymal tumor, malignant</li> <li>• Synovial sarcoma (NOS, spindle cell, biphasic)</li> <li>• Epithelioid sarcoma</li> <li>• Alveolar soft part sarcoma</li> <li>• Clear cell sarcoma of soft tissue</li> <li>• Extraskeletal myxoid chondrosarcoma</li> <li>• Extraskeletal Ewing sarcoma</li> <li>• Desmoplastic small round cell tumor</li> <li>• Extrarenal rhabdoid tumor</li> <li>• Perivascular epithelioid cell tumor (PEComa), NOS</li> <li>• Intimal sarcoma</li> </ul>
<b>Undifferentiated/ unclassified sarcoma</b>	<ul style="list-style-type: none"> <li>• Undifferentiated (spindle cell sarcoma, pleomorphic sarcoma, round cell sarcoma, epithelioid sarcoma, NOS)</li> </ul>

## Risk factors

Anything that increases your chances of cancer is called a risk factor. Risk factors can be activities that people do, things you have contact with in the environment, or traits passed down from parents to children through genes (inherited). Genes are coded instructions that tell your cells what to do and what to become.

Certain genetic syndromes may put someone at risk for developing soft tissue sarcoma. Share what you know about your family history with your doctor.

Some syndromes that increase the risk for soft tissue sarcoma include:

- Li-Fraumeni syndrome
- Familial adenomatous polyposis (FAP) and Gardner's syndrome
- Carney-Stratakis syndrome
- Hereditary retinoblastoma
- Neurofibromatoses
- *BRCA2* gene mutations

Your health care provider might refer you for genetic testing to learn more about your cancer. A genetic counselor will speak to you about the results. A genetic counselor is an expert who has special training in genetic diseases.

## Review

- Sarcomas are a large but rare group of cancers that start in the connective tissue of the body.
- Sarcomas start in cells that make up bones or soft tissue.
- Soft tissue includes fat, muscles, nerves, tendons, and blood and lymph vessels. Soft tissue sarcoma starts in the mesenchymal cells that develop into soft tissue.
- There are over 50 types of soft tissue sarcoma that can be found anywhere in the body.
- Diagnosis and treatment are based on the type and location of the soft tissue sarcoma.
- Certain genetic syndromes may put you at risk for developing soft tissue sarcoma. Therefore, you might be referred to genetic counseling.

# 2

## Testing for soft tissue sarcoma

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14	Test results
14	General health tests
15	Imaging tests
17	Tissue tests
18	Genetic tests
19	Cancer staging
20	Review



Treatment planning starts with testing. Accurate testing is needed to diagnose and treat soft tissue sarcoma. A biopsy is recommended before starting treatment. This chapter presents an overview of the tests you might receive and what to expect.

## Test results

Results from blood tests, imaging studies, and biopsy will determine your treatment plan. It is important you understand what these tests mean. Ask questions and keep copies of your test results. Online patient portals are a great way to access your test results.

Whether you are going for a second opinion, test, or office visit, keep these things in mind:

- Bring someone with you to doctor visits. Encourage this person to ask questions and take notes.
- Write down questions and take notes during appointments. Don't be afraid to ask your care team questions. Get to know your care team and let them get to know you.
- Get copies of blood tests, imaging results, and reports about the specific type of cancer you have. It will be helpful when getting a second opinion.
- Organize your papers. Create files for insurance forms, medical records, and test results. You can do the same on your computer.

- Keep a list of contact information for everyone on your care team. Add it to your binder or notebook. Hang the list on your fridge or keep it by the phone.

For recommended tests, [see Guide 3](#).

## General health tests

### Medical history

A medical history is a record of all health issues and treatments you have had in your life. Be prepared to list any illness or injury and when it happened. Bring a list of old and new medicines and any over-the-counter medicines, herbs, or other supplements you take. Tell your doctor about any symptoms you have. A medical history will help determine which treatment is best for you.

### Family history

Some cancers and other diseases can run in families. Your doctor will ask about the health history of family members who are blood relatives. This information is called a family history. You can ask family members about their health issues like heart disease, cancer, and diabetes, and at what age they were diagnosed.

### Physical exam

A physical exam is a study of your body. Doctors should perform a thorough physical exam along with a complete health history. A doctor will check your body for signs of disease.



A health care provider may:

- Check your temperature, blood pressure, pulse, and breathing rate
- Weigh you
- Listen to your lungs and heart
- Look in your eyes, ears, nose, and throat
- Feel and apply pressure to parts of your body to see if organs are of normal size, are soft or hard, or cause pain when touched. Tell your doctor if you feel pain.
- Feel for enlarged lymph nodes in your neck, underarm, and groin. Tell the doctor if you have felt any lumps or have any pain.
- Measure the size of a tumor that is on the surface or close to the skin

## Imaging tests

Imaging tests take pictures of the inside of your body. These tests are used to find and treat soft tissue sarcoma. Imaging tests show the primary tumor, or where the cancer started, and look for cancer in other parts of the body.

A radiologist, an expert who looks at test images, will review test images and write a report. The radiologist will send this report to your doctor who will discuss the results with you. Feel free to ask as many questions as you like.

### Guide 3

#### Tests for soft tissue sarcoma

<b>Needed</b>	Before starting therapy, all patients should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma
	Medical history and physical exam
	Imaging of primary tumor and as needed for lesions that could be cancer
	Carefully planned needle biopsy (preferred) or incisional biopsy based on adequate imaging
	Chest imaging
<b>Useful in some cases (different subtypes have a tendency to spread to various locations)</b>	Other imaging as needed
	Consider more genetic testing in those with a personal or family history of Li-Fraumeni syndrome
	Other testing as needed for those with neurofibromatosis type 1

**MRI scan**

A magnetic resonance imaging (MRI) scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays.

Before an MRI, you may be given a contrast. Contrast material is used to improve the pictures inside the body. Contrast materials are not dyes, but substances that help certain areas in the body stand out. Contrast is used to make the pictures clearer.

Tell your doctors if you have had bad reactions to contrast in the past. This is important. You might be given medicines, such as Benadryl® and prednisone, for an allergy to contrast. Contrast might not be used if you have a serious allergy or if your kidneys aren't working well.

An MRI with or without contrast is recommended to provide details about the size of the tumor and its location (proximity) to nearby internal structures such as organs, nerves, and blood vessels.

**CT scan**

A computed tomography (CT) scan uses x-rays and computer technology to take pictures of the inside of the body. It takes many x-rays of the same body part from different angles. All the images are combined to make one detailed picture.

A CT scan of your chest, abdomen, and/or pelvis may be one of the tests to look for cancer. It might be given in addition to an MRI scan. In most cases, contrast will be used.

**PET scan**

Positron emission tomography (PET) scans use a radioactive drug called a tracer to find disease and take three-dimensional or real-looking pictures. A tracer is a substance put in your body to see how cancer is growing and where it is in the body. Cancer cells show up as bright spots on PET scans. Not all bright spots are cancer. A PET scan combined with a CT (PET/CT) may be used to look for small tumors in soft tissue and to see if cancer has spread.

**X-ray**

An x-ray uses low-dose radiation to take one picture at a time. A tumor changes the way radiation is absorbed and will show up on the x-ray. A chest x-ray may be used with other initial tests when sarcoma is first suspected or found. It may also be done to check treatment results.

**Ultrasound**

An ultrasound uses high-energy sound waves to form pictures of the inside of the body. Ultrasound is good at showing small areas of cancer that are superficial (on the surface of the body).

**Angiogram**

An angiogram is an x-ray or computer image (CT scan or MRI) of the blood vessels. It is used to look for a blockage or tumor that might affect blood flow. This type of test uses contrast put into a catheter (thin, flexible tube). The catheter is inserted into an artery or vein.

## Tissue tests

A biopsy, or tissue sample, is needed to diagnose soft tissue sarcoma. A biopsy is recommended before starting treatment.

A biopsy is the removal of tissue or group of cells by a surgeon. A biopsy looks for cancerous cells. After imaging tests, your doctor will order a biopsy to learn more about your type of soft tissue sarcoma. Sometimes, a sample of tissue from the biopsy does not have enough cells to check for cancer. The tissue could also be abnormal but not cancer. If this happens, you may have another biopsy.

A pathologist is an expert who will test the biopsy for cancer and write a report called a pathology report. The pathologist may perform other tests to see if the tumor cells have specific genes or proteins. This information will help choose the best treatment plan for your type of soft tissue sarcoma. Because not all pathologists are familiar with every type of soft tissue sarcoma, review at an experienced center is recommended. Ask questions about your biopsy results and what it means for your treatment.

There are different types of biopsies. Some biopsies are guided using imaging, such as an ultrasound or CT. The primary or main tumor is biopsied first. Other tumors or tumors in different areas may also be biopsied.

Types of possible biopsies include:

- ▶ **Fine-needle aspiration (FNA)** uses a thin needle to remove a sample of tissue or fluid. An ultrasound may guide the FNA.
- ▶ **Core needle biopsy** removes tissue samples with a wide, hollow needle. This is usually the preferred biopsy for soft tissue sarcoma.
- ▶ **Incisional biopsy** removes a small amount of tissue through a cut in the skin or body.

Other image-guided needle biopsies may be needed for tumors in the chest, abdomen, pelvis, or other deep areas.

## Genetic tests

Anything that increases your chances of cancer is called a risk factor. Risk factors can be activities that people do, things you have contact with in the environment, or traits passed down from parents to children through genes (inherited). Genes are coded instructions that tell your cells what to do and what to become. An abnormal change in these instructions—called a gene mutation—can cause cells to grow and divide out of control. Gene mutations may be associated with certain types of sarcoma. More information can be found in the back of this book; [see Guide 38](#).

Certain genetic syndromes may put someone at risk for developing soft tissue sarcoma. Share what you know about your family history with your doctor.

Some syndromes that increase the risk for soft tissue sarcoma include:

- Li-Fraumeni syndrome
- Familial adenomatous polyposis (FAP) and Gardner's syndrome
- Carney-Stratakis syndrome
- Hereditary retinoblastoma
- Neurofibromatosis
- *BRCA2* gene mutations

Your health care provider might refer you for genetic testing to learn more about your cancer. A genetic counselor will speak to you about the results. A genetic counselor is an expert who has special training in genetic diseases.

## Recommendations

NCCN recommendations are described next.

### Li-Fraumeni syndrome

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

### FAP

People with FAP have an adenomatous polyposis coli (*APC*) gene mutation. This syndrome causes colon polyps that may become colon cancer. A polyp is a growth from the inner wall of the digestive tract. FAP has a related syndrome called Gardner's syndrome. Both are associated with desmoid tumors. Therefore, if you are diagnosed with a desmoid tumor, then your family history should be looked at closely. A colonoscopy should be considered.

### *SDH* gene mutation

Succinate dehydrogenase (SDH) is a protein within cells that converts energy. If you are diagnosed with a gastrointestinal stromal tumor (GIST) that doesn't have *KIT* or *PDGFRA* gene mutation, then additional genetic testing should be considered to look for SDH.

## Cancer staging

The American Joint Committee on Cancer (AJCC) created a way to determine how much cancer is in your body and where it is located. This is called staging. Based on testing, your cancer will be assigned a stage. Staging is needed to make treatment decisions.

Not all soft tissue sarcomas are staged the same way. Some do not use a staging system.

### TNM scores

The TNM staging system is used for soft tissue sarcomas in the limbs and outer torso. The tumor, node, metastasis (TNM) system is used to stage many soft tissue sarcomas. In this system, the letters T, N, and M describe different areas of cancer growth. Based on cancer test results, your doctor will assign a score or number to each letter. The higher the number, the larger the tumor or the more the cancer has spread. These scores will be combined to assign the cancer a stage. A TNM example might look like this: T2, N0, M0.

- **T (tumor)** - Size of the main (primary) tumor
- **N (node)** - If cancer has spread to nearby (regional) lymph nodes. Lymph node involvement is uncommon in most soft tissue sarcomas.
- **M (metastasis)** - If cancer has spread to distant parts of the body or metastasized

### Grade

Another factor used in staging is the tumor grade. Grade describes how abnormal the tumor cells look under a microscope and how quickly these cells are likely to grow and spread. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. The letter G stands for the grade. GX means the grade can't be determined, followed by G1, G2, and G3. G3 is the highest grade.

The French Federation of Cancer Centers Sarcoma Group (FNCLCC) is used to grade tumors.

There are 3 categories for grade:

- **Differentiation** - how much the cancer cells look like normal cells
- **Mitosis** - how fast the cancer cells make copies of themselves
- **Necrosis** - how much dead tissue is in the tumor

### Stages

Number stages range from stage 1 to stage 4, with 4 being the most advanced. Doctors write these stages as stage I, stage II, stage III, and stage IV. These stages are used for sarcomas in the torso, limbs, retroperitoneum, and GIST.

Not every soft tissue sarcoma is given a stage. Staging is based on the regular ability of a tumor to metastasize. Soft tissue tumors like desmoid tumors are locally aggressive, but not metastatic. This means the tumor tends to return near the same place, but not distant sites.

## Review

- Tests are used to find cancer, plan treatment, and check how well treatment is working.
- Imaging tests take pictures of the inside of your body.
- MRI and CT scans provide details about the size of the tumor and its location (proximity) to nearby internal structures such as organs, nerves, and blood vessels.
- A biopsy removes tissue or fluid from your body to diagnose (confirm) cancer. A biopsy is recommended before starting treatment.
- A core needle biopsy is usually the preferred type of biopsy for soft tissue sarcoma.
- Your health care provider might refer you for genetic testing to learn more about your cancer.
- Based on testing, your soft tissue sarcoma might be assigned a TNM score, grade, and/or stage. Staging is used to make treatment decisions.



### Create a medical binder

A medical binder or notebook is a great way to organize all of your records in one place.

- Make copies of blood tests, imaging results, and reports about your specific type of cancer. It will be helpful when getting a second opinion.
- Choose a binder that meets your needs. Consider a zipper pocket to include a pen, small calendar, and insurance cards.
- Create folders for insurance forms, medical records, and tests results. You can do the same on your computer.
- Use online patient portals to view your test results and other records. Download or print the records to add to your binder.
- Organize your binder in a way that works for you. Add a section for questions and to take notes.
- Bring your medical binder to appointments. You never know when you might need it!

# 3

## Treatment options

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22	Surgery
23	Radiation therapy
25	Other local treatments
26	Systemic therapy
27	Clinical trials
28	Treatment team
29	Review



Treatment options depend on the location and type of soft tissue sarcoma. Learn about the treatments used to treat soft tissue sarcoma. Not everyone will receive the same treatment. Discuss with your doctor which treatment might be best for you.

Treatment can be local, systemic, or a combination of both.

- Local therapy focuses on a certain area. It includes surgery and radiation therapy.
- Systemic therapy works throughout the body. It includes chemotherapy, targeted therapy, and immunotherapy.

## Surgery

Surgery is a form of local treatment. It is an operation or procedure to remove cancer from the body. This is only one part of a treatment plan.

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

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

When surgery involves your limbs (an arm or a leg), the goal is to have a functional or working limb. Radiation may be given before or after surgery. This is to prevent the need for amputation.

### Biopsy

A biopsy to diagnose and grade a sarcoma is preferred before treatment. Biopsy should be carried out by an experienced surgeon (or radiologist) and may be an incisional or needle biopsy. A core needle biopsy is preferred.

### Tumor resection

Imaging tests will be used to see if your cancer is resectable (can be removed completely with surgery) or unresectable (cannot be removed with surgery).

The goal of surgery or tumor resection is to remove all of the cancer. To do so, the tumor is removed along with some normal-looking tissue around its edge. The normal-looking tissue is called the surgical margin. A clear or negative margin (R0) is when no cancerous cells are found in the tissue around the edge of the tumor. In an R1 positive margin, the surgeon removes all of the visible tumor, but the microscopic margins are still positive for tumor cells. In an R2 positive margin, the surgeon does not or is unable to remove all of the visible tumor.

A negative margin (R0) is the best result. Your surgeon will look carefully for cancer not only along the surgical margin, but in other nearby areas. It is not always possible to find all of the cancer.

Sometimes, surgeons can't safely remove the sarcoma with a cancer-free margin. If this is



expected, your surgeon will place clips in your body after removing the tumor. The clips will help doctors know where to give you radiation therapy. You might have more than one surgery. You might also have a wound drain to prevent fluid from collecting in the body after surgery.

### Limb-sparing

For sarcomas in the limbs, the goal of surgery, whenever possible, is to preserve or maintain limb function. This is called limb-sparing surgery.

Rehabilitation, such as physical and occupational therapy, will likely be part of a limb-sparing treatment plan. This may include occupational therapy to help with daily life skills or physical therapy to help your body move and function.

### Amputation

Amputation is the removal of a limb or other body part. A surgeon who is an expert in soft tissue sarcoma should be consulted before amputation. Rehabilitation, such as physical and occupational therapy, will be part of this treatment. Seek a peer support group to connect with others who have experienced the loss of a limb. This will aid in your recovery and help you to maintain an active lifestyle.

## Radiation therapy

Radiation therapy is a form of local treatment that has many uses in the treatment of soft tissue sarcoma.

Radiation therapy uses high-energy radiation from x-rays, gamma rays, protons, and other

sources to kill cancer cells and shrink tumors. It is given over a certain period of time. Radiation therapy may be given alone or before or after surgery to treat or slow the growth of cancer. It may be used as supportive care to help ease pain or discomfort.

Radiation may be given:

- As the primary (first) treatment for sarcoma
- Before surgery, called neoadjuvant therapy, to shrink the tumor before surgery. The concern with radiation therapy given before surgery is it can slow wound healing.
- During surgery, called intraoperative radiation therapy (IORT)
- After surgery, called adjuvant treatment, to kill any cancerous cells that remain. The concern with radiation therapy given after surgery is bone fracture. This can happen at a later time. It is caused by the higher dose of radiation used in adjuvant treatment.

There are different ways to give radiation. Which method you receive will depend on the size and type of sarcoma and the purpose of radiation therapy.

There are 2 main types of radiation treatment:

- **External beam radiation therapy (EBRT)** uses a machine outside of the body to aim radiation at the tumor(s).
- **Internal radiation** is placed inside the body as a solid like seeds. This is called brachytherapy.

**EBRT**

There is more than one type of EBRT used in the treatment of soft tissue sarcoma. These allow for safer, higher doses of radiation.

Types of EBRT that may be used to treat your cancer include:

- ▶ **Stereotactic body radiation therapy (SBRT)** uses high-energy radiation beams to treat cancers.
- ▶ **Proton beam radiation therapy** uses streams of particles called protons to kill tumor cells.
- ▶ **Three-dimensional conformal radiation therapy (3D-CRT)** uses computer software and CT images to aim beams that match the shape of the tumor.
- ▶ **Intensity-modulated radiation therapy (IMRT)** uses small beams of different strengths to match the shape of the tumor.
- ▶ **Image-guided radiation therapy (IGRT)** uses a computer to create a picture of the tumor. This helps guide the radiation beam during treatment. It is used with IMRT and 3D-CRT. Tumors can shift slightly within the body and can change shape and size between and during treatment visits. Because of this, IGRT can improve how well 3D-CRT and IMRT target the tumor.
- ▶ **Intraoperative radiation therapy (IORT)** uses radiation treatment aimed directly at the tumor during surgery.

**Radiation therapy**

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



## Brachytherapy

Brachytherapy is another type of radiation therapy. In this treatment radiation is placed inside or next to the tumor. Brachytherapy may be used alone or combined with EBRT. You might hear it called brachy (said “BRAY-key”), for short.

There are 2 types of brachytherapy used to treat soft tissue sarcoma:

- Low dose-rate (LDR) brachytherapy
- High dose-rate (HDR) brachytherapy

### LDR brachytherapy

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

The seeds usually consist of either radioactive iodine or palladium. They will stay in your tumor and give a low dose of radiation for a few months. The radiation will travel a very short distance. This will allow for a large amount of radiation within a small area while sparing nearby healthy tissue. Over time, the seeds will stop radiating, but will stay in your body (permanent).

### HDR brachytherapy

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

### Brachytherapy boost

Brachytherapy used with EBRT is called a brachytherapy boost, or brachy boost for short.

## Other local treatments

### Ablation

Ablation is a type of local treatment that uses extreme cold or extreme heat to destroy cancer cells.

It can destroy small tumors with little harm to nearby tissue.

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

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

Both types of ablation use a special needle, called a probe, which is inserted into the tumor. With cryotherapy, a medical gas is passed through the probe to cause below-freezing temperatures. This freezes the tumor to destroy it. With radiofrequency ablation, the probe emits radio waves to heat the tumor and destroy it.

The probe can be guided into place with a CT scan, ultrasound, or other imaging tests. The probe 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.

## Systemic therapy

A cancer treatment that affects the whole body is called systemic therapy. Chemotherapy used to be the most common type of systemic therapy. Now, there are other cancer treatments like targeted therapy and immunotherapy. Each works differently to shrink the tumor and prevent recurrence. Systemic treatments may be used alone or together.

Systemic therapies that might be used include:

- **Chemotherapy** – attacks rapidly dividing cells in the body
- **Targeted therapy** – focuses on specific or unique feature of cancer cells
- **Immunotherapy** – uses your body's natural defenses to find and destroy cancer cells

### Chemotherapy

Chemotherapy is a type of drug therapy used to treat cancer. Chemotherapy kills fast-growing cells throughout the body, including cancer cells and normal cells.

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

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

### Did you know?

The terms “chemotherapy” and “systemic therapy” are often used interchangeably, but they are not the same. Chemotherapy, targeted therapy, and immunotherapy are all types of systemic therapy.

Most chemotherapy is given in cycles of treatment days followed by days of rest. This allows the body to recover before the next cycle. Cycles vary in length depending on which drugs are used. The number of treatment days per cycle and the total number of cycles given also vary.

### Chemoradiation

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

### Targeted therapy

Targeted therapy is a form of systemic treatment that works throughout your body. It is drug therapy that focuses on specific or unique features of cancer cells.

Targeted therapies seek out how cancer cells grow, divide, and move in the body. These drugs stop the action of molecules that help cancer cells grow and/or survive. 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. Others target hormones.

Targeted therapy isn't used for every sarcoma. It might be used alone or with chemotherapy.

The following are some targeted therapies that might be used to treat soft tissue sarcoma:

- Everolimus
- Temsirolimus
- Sorafenib
- Sunitinib
- Pazopanib
- Bevacizumab

### Immunotherapy

The immune system is the body's natural defense against infection and disease. It is a complex network of cells, tissues, and organs. 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.

Pembrolizumab, ipilimumab, and nivolumab are types of immunotherapy.

## Clinical trials

Clinical trials study how safe and helpful tests and treatments are for people. Clinical trials find out how to prevent, diagnose, and treat a disease like cancer. Because of clinical trials, doctors find safe and helpful ways to improve your care and treatment of cancer.

Clinical trials have 4 phases.

- **Phase I trials** aim to find the safest and best dose of a new drug. Another aim is to find the best way to give the 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 a standard treatment.
- **Phase IV trials** test drugs approved by the U.S. FDA (Food and Drug Administration) to learn more about side effects with long-term use.

To join a clinical trial, you must meet the conditions of the study. Patients in a clinical trial often are alike in terms of their cancer and general health. This helps to ensure that any change is from the treatment and not because of differences between patients.

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

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. Discuss the risks and benefits of joining a clinical trial with your care team. Together, decide if a clinical trial is right for you.

NCCN experts encourage patients to join a clinical trial, when possible.

## Treatment team

Treating soft tissue sarcoma takes a team approach. NCCN recommends that treatment decisions involve a multidisciplinary team or a team of doctors from different fields of medicine who have knowledge (expertise) and experience with your type of soft tissue sarcoma. If you have sarcoma, it is important that the experts meet before your treatment starts.

Some members of your care team will be with you throughout cancer treatment, while others will only be there for parts of it. Get to know your care team and let them get to know you.

- Your primary care doctor handles medical care not related to your cancer. This person can help you express your feelings about treatments to your cancer care team.

- **A pathologist** reads tests and studies the cells, tissues, and organs removed during a biopsy or surgery.
- **A diagnostic radiologist** reads the results of x-rays and other imaging tests.
- **A surgical oncologist** performs operations to remove cancer.
- **A medical oncologist** treats cancer in adults using systemic therapy. Often, this person will lead the overall treatment team and keep track of tests and exams done by other specialists.
- **Advanced practice providers** are an important part of any team. These are registered nurse practitioners and physician assistants who monitor your health and provide care.
- **Oncology nurses** provide your hands-on care, like giving systemic therapy, managing your care, answering questions, and helping you cope with side effects.

Depending on your diagnosis, your team might include:

- **An anesthesiologist** who gives anesthesia, a medicine so you do not feel pain during surgery or procedures
- **An interventional radiologist** who performs needle biopsies of tumors and sometimes performs ablation therapies
- **A radiation oncologist** who prescribes and plans radiation therapy to treat cancer
- **A gastroenterologist** who is an expert in digestive diseases
- **A thoracic surgeon** who performs operations within the chest

- **An orthopedic surgeon** who performs operations that involve bones, joints, tendons, ligaments, and muscles
- **A plastic surgeon** who performs operations to improve function and appearance
- **An occupational therapist** who helps people with the tasks of daily living
- **A physical therapist** who helps people move with greater comfort and ease
- **A dietitian or nutritionist** who gives food advice and helps plan menus to meet your nutrition needs during cancer treatment

You know your body better than anyone. Help other team members understand:

- How you feel
- What you need
- What is working and what is not

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

## Review

- Local treatments for soft tissue sarcoma include surgery, radiation therapy, ablation, and embolization.
- Surgery removes the tumor along with some normal-looking tissue around its edge called a surgical margin.
- Radiation kills cancer cells or stops new cancer cells from being made.
- Systemic treatments treat cancer throughout the body.
- Systemic treatments include drug treatments such as chemotherapy, targeted therapy, and immunotherapy.
- Chemotherapy kills fast-growing cells throughout the body, including cancer cells and normal cells.
- Targeted therapies seek out how cancer cells grow, divide, and move in the body.
- Immunotherapy uses your body's natural defenses to find and destroy cancer cells.
- A clinical trial is a type of research that studies a treatment to see how safe it is and how well it works.

# 4

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

31	Overview
32	Tests
33	Stages
36	Stage 1
38	Stage 2 and 3
45	Stage 4
47	Recurrence
49	Review





This chapter includes treatment options for sarcomas in the arms, legs, outer torso, or head and neck. Cancer staging is how doctors rate and describe the amount of cancer in your body. Doctors use staging to plan treatment. Together, you and your doctor will choose a treatment plan that is best for you.

## Overview

There are over 50 types of soft tissue sarcoma. Any number of these subtypes can be located in the extremities such as the arms and legs, outer (superficial) torso, or head and neck. Therefore, testing is very important to diagnose and treat tumors found in these areas. Treatment is based on cancer stage, the type of tumor(s), and the location of the tumor(s).

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

- ▶ **Alveolar soft part sarcoma** begins in the soft tissue that surrounds organs and other tissues. It usually occurs in the thighs, but can be found in the arms, legs, or torso.
- ▶ **Angiosarcoma** begins in the cells that line blood vessels or lymph vessels.
- ▶ **Epithelioid sarcoma** begins as a slow-growing, firm lump in the deep soft tissue or skin of the arms, hands, or fingers. It may also occur in the legs, torso, or head and neck. It is usually painless, but it has a tendency to spread and to return after treatment.
- ▶ **Leiomyosarcoma** is cancer of the smooth muscle cells that can arise almost anywhere in the body.
- ▶ **Myxoid/round cell liposarcoma** begins in fat cells. It usually occurs in the thigh, but can be found in the outer torso and buttocks.
- ▶ **Well-differentiated liposarcoma** cells look like fat cells under a microscope. It tends to grow more slowly than poorly differentiated or undifferentiated cancer cells.
- ▶ **Dermatofibrosarcoma protuberans** with fibrosarcomatous changes and/or malignant changes start as a hard nodule that grows slowly. It is usually found in the deeper layer of tissue under the skin. These tumors are related to giant cell fibroblastomas.

## Tests

You will have a variety of tests based on the suspected type of soft tissue sarcoma and to rule out desmoid tumors and other lesions. Your doctors will take a medical history, complete a physical exam of your body, and

order imaging tests. A biopsy should be done to confirm cancer and determine the specific type of soft tissue sarcoma you have. A core needle biopsy is preferred. For recommended tests, [see Guide 4](#).

### Guide 4

#### Tests for sarcomas in the limbs, outer torso, or head and neck

##### Needed

Before starting therapy, all patients should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma

Medical history and physical exam

Imaging of primary tumor using MRI with or without contrast. A CT with contrast might be used with the MRI

Carefully planned needle biopsy (preferred) or incisional biopsy based on adequate imaging

Chest imaging such as x-ray or CT without contrast (preferred)

##### Useful

Other imaging, such as PET/CT for cancer stage, prognosis, and grade

Other imaging, such as angiogram and x-ray

Consider CT of abdomen/pelvis for myxoid/round cell liposarcoma, epithelial sarcoma, angiosarcoma, and leiomyosarcoma

Consider MRI of total spine for myxoid/round cell liposarcoma

Consider central nervous system imaging with MRI or CT for alveolar part sarcoma and angiosarcoma

Consider CT of pelvis for lower-limb well-differentiated liposarcoma

Consider more genetic testing in those with personal or family history of Li-Fraumeni syndrome

Other testing for those with neurofibromatosis type 1

Soft tissue sarcomas in the arms, legs, outer torso, or head and neck can spread to distant sites in the body (metastasize). You might have imaging tests that will look for cancer that has metastasized to the lungs, brain, or spine. Given there is a risk of cancer spreading to the lungs, CT scan of the chest with contrast is preferred for accurate staging.

Other imaging will be considered based on the subtype:

- Alveolar soft part sarcoma and angiosarcoma can spread to the brain and central nervous system. Therefore, a brain MRI and MRI or CT of the total spine will be considered.
- A CT of the pelvis will be considered for lower-limb well-differentiated sarcoma.
- A CT of the abdomen/pelvis will be considered for angiosarcoma, epithelioid sarcoma, leiomyosarcoma, and myxoid/round cell liposarcoma.
- Myxoid/round cell liposarcoma has a high risk of spreading to the spine. Therefore, an MRI of the total spine will be considered.

Treatment planning is based on test results. Testing is needed to diagnose (confirm) soft tissue sarcoma. Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating your type of soft tissue sarcoma.

## Stages

### Head and neck

Head and neck soft tissue sarcomas are similar to sarcomas in other parts of the body. However, sarcomas in the head and neck can be more challenging because of the location. The tumor is likely to be close to bones, muscles, and nerves making surgery more difficult.

Areas of the head and neck that could be affected include:

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

TNM staging and grade are used to diagnose and treat soft tissue sarcomas of the head and neck. These scores are not divided into stages like soft tissue sarcomas of the limbs and outer torso. TNM scores and grades for soft tissue sarcomas of the head and neck are described next.

### T = Tumor

The primary tumor size is measured in centimeters (cm). A large pea is 1 cm. A golf ball is 4 cm.

- **T1** Tumor is 2 cm or less
- **T2** Tumor is 2.1 cm to 4 cm
- **T3** Tumor is more than 4 cm
- **T4a** Tumor has invaded nearby structures such as the skull and muscles
- **T4b** Tumor has invaded the brain, carotid artery, muscles, or central nervous system

**N = Node**

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are those located near the tumor. Certain subtypes are more likely to spread to lymph nodes. These include epithelioid sarcoma, clear cell sarcoma, angiosarcoma, and rhabdomyosarcoma.

- **N0** means there is no regional lymph node metastasis or unknown
- **N1** means regional lymph node metastasis is found

**M = Metastatic**

Cancer that has spread to distant parts of the body is shown as M1. The most common site for metastasis is the lung.

- **M0** means no distant metastasis
- **M1** means distant metastasis is found

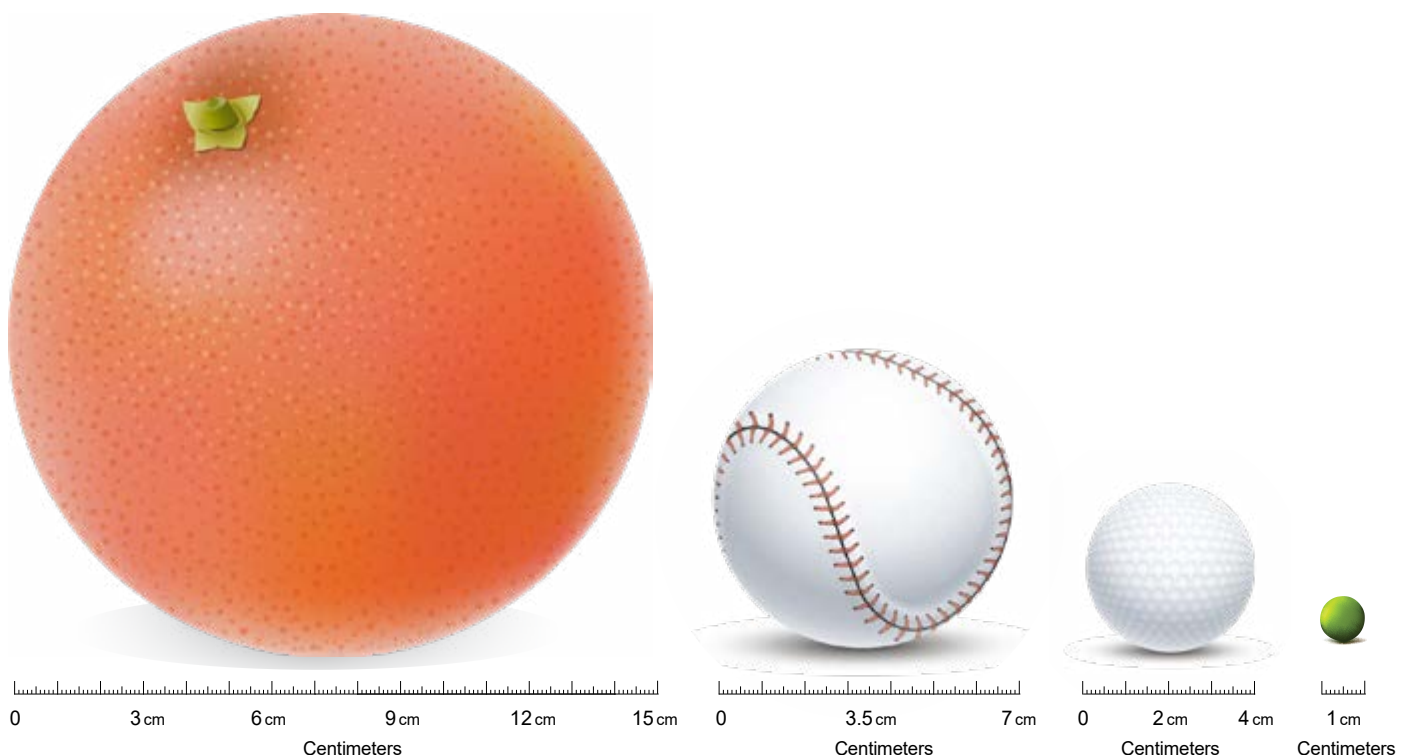
**G = Grade**

Grade describes how abnormal the tumor cells look under a microscope and how quickly these cells are likely to grow and spread. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. G3 is the highest grade.

- **GX** means grade cannot be assessed

**Soft tissue sarcoma tumors are measured in centimeters**

A grapefruit is 15 cm, a baseball is 7 cm, a golf ball is 4 cm, and a large pea is 1 cm.



- **G1** is a score of 2 or 3
- **G2** is a score of 4 or 5
- **G3** is a score of 6, 7, or 8

### Outer torso and extremities

The TNM staging system is used for soft tissue sarcomas in the limbs and outer torso. The limbs include the arms, legs, and thighs. Outer torso includes the chest, back, buttocks, and abdomen. Staging is based on imaging and biopsy results. For stages, [see Guide 5](#).

### T = Tumor

The primary tumor size is measured in centimeters (cm). A golf ball is 4 cm. A baseball is 7 cm. A grapefruit is 15 cm.

- **T1** Tumor is 5 cm or less
- **T2** Tumor is 5.1 cm to 10 cm
- **T3** Tumor is 10.1 cm to 15 cm
- **T4** Tumor is more than 15 cm

## Guide 5

### Cancer stages for soft tissue tumors in the limbs and outer torso

Stage 1	Stage 1A	• T1, N0, M0, G1 or GX
	Stage 1B	• T2, N0, M0, G1 or GX • T3, N0, M0, G1 or GX • T4, N0, M0, G1 or GX
Stage 2	• T1, N0, M0, G2 or G3	
Stage 3	Stage 3A	• T2, N0, M0, G2 or G3
	Stage 3B	• T3, N0, M0, G2 or G3 • T4, N0, M0, G2 or G3
Stage 4	• Any T, N1, M0, Any G • Any T, Any N, M1, Any G (metastatic)	

**N = Node**

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are those located near the tumor.

- **N0** means no regional lymph node metastasis or unknown
- **N1** means regional lymph node metastasis is found

**M = Metastatic**

Cancer that has spread to distant parts of the body is shown as M1. The most common site for metastasis is the lung.

- **M0** means no distant metastasis
- **M1** means distant metastasis is found

**G = Grade**

Grade describes how abnormal the tumor cells look under a microscope and how quickly these cells are likely to grow and spread. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. G3 is the highest grade.

- **GX** means grade cannot be assessed
- **G1** is a score of 2 or 3
- **G2** is a score of 4 or 5
- **G3** is a score of 6, 7, or 8

**Stage 1****Any T, N0, M0, G1 or GX**

In stage I, the tumor can be any size (T), and it has not spread to nearby lymph nodes (N0) or metastasized to distant sites (M0). The grade must be G1 or GX. Stage 1 is further grouped into stage 1A and stage 1B.

Surgery to remove the tumor is called resection. It is the primary or main treatment for stage 1. For stage 1, the intent of surgery is to cure the cancer. Surgery has risk. Those with some types of soft tissue sarcoma have a greater risk of complications from surgery. Also, sarcomas found in areas of the face, head, and neck might be more difficult to remove. Ask your doctor about the risks of surgery, what will be removed, what this means in terms of your recovery, and what to expect.

Removing the sarcoma with a cancer-free surgical margin (R0) is the goal. However, to avoid cutting nerves and blood vessels, your surgeon may not be able to remove a large enough surgical margin. In this case, there is a higher chance that cancer cells will be left behind. A pathologist will look at a sample of the surgical margin under a microscope to look for cancer cells. If cancer is found, you may have a second surgery or radiation therapy.

Treatment options are based on if the:

- Surgical margin is 1 cm or less
- Fascia (layer of connective tissue) was cut

Recommendations for more treatment are based on:

- The size of the margin
- If the surgical cut went deep into the fascia

### **Sarcoma was resected**

If your soft tissue sarcoma was completely removed with surgery (resected) and a negative surgical margin was achieved (R0), then you can start follow-up care.

### **Surgical margin was small**

If not enough tissue around the sarcoma was removed (resected), then your next treatment options are:

- Re-resection (another surgery)
- Observation for stage 1A tumors
- Consider radiation therapy

The results of primary treatment are important. After surgery, your doctors will assess if you need further treatment. Based on the margins this could mean more surgery for a positive margin that does not include bone, major blood vessels, or nerves. Doctors may suggest observation after primary treatment. Observation is a period of scheduled follow-up testing to watch for signs of cancer spread (metastasis) or return (recurrence). Observation is an option for stage 1A tumors. Another option is radiation therapy to lower the chances of cancer returning. Radiation therapy is a very good option for stage 1B tumors. All options carry risk.

After treatment is complete, you can start follow-up care.

### **Follow-up care**

Follow-up care will begin when treatment is complete. Tests will look for signs of cancer that has returned called recurrence. Follow-up tests can help find cancer early.

Follow-up tests include a medical history, physical exam, and imaging tests. You will have a medical history and physical exam every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years, followed by once a year. You should be evaluated for rehabilitation, such as occupational therapy to help with daily life skills or physical therapy to help your body move and function.

If 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 if the risk of recurrence is high. Ask your doctor about what follow-up care you may need.

Your doctor will consider the tests listed in [Guide 6](#).

## Stage 2 and 3

Treatment options for sarcoma are based on whether surgery can be done. A tumor that can be removed with surgery is called resectable. Surgery might not be the first or best treatment option. It might be because the tumor is too large. Sometimes, removing a sarcoma would limit the use of a limb or other body part. Surgery could cause health-related problems. Therefore, treatment is grouped into resectable with good function, resectable with poor function, and unresectable.

### Resectable with good function

If a tumor can be safely removed with surgery and you will have good use of the limb or area, then it is considered resectable. Surgery will

be the primary treatment. You might have treatment before and/or after surgery. For stage 2 and 3 sarcomas that are resectable with good function, [see Guide 7](#).

### Surgery

All options include surgery to remove the tumor. Multiple treatments can be combined with surgery and may be given before surgery (preoperative or neoadjuvant) or after surgery (postoperative or adjuvant).

The following options include surgery:

- Radiation therapy
- Chemoradiation
- Systemic therapy

## Guide 6

### Follow-up testing to consider: Stage 1

Chest imaging every 6 to 12 months. X-ray or CT is preferred.

Imaging of the primary site using MRI with or without contrast and/or CT with contrast

Ultrasound of small lesions near the surface of the skin done by someone experienced in muscle/skeletal disease

CT of abdomen/pelvis for myxoid/round cell liposarcoma, epithelial sarcoma, angiosarcoma, and leiomyosarcoma

MRI of total spine for myxoid/round cell liposarcoma

Central nervous system imaging with MRI or CT for alveolar part sarcoma and angiosarcoma

CT of pelvis for lower-limb well-differentiated liposarcoma



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

A PET/CT may be used to find out if the cancer has responded to systemic therapy.

### Radiation therapy

Radiation therapy before surgery may reduce the tumor size for surgery and lower the chances of the cancer returning. It may also improve how well your limb works after surgery. However, radiation will likely slow healing of the surgical wound. Before having surgery, you may get another imaging test. This test

will assess the tumor and rule out metastatic disease.

### Systemic therapy

The systemic therapy used will depend on the subtype of soft tissue sarcoma.

- If you have a non-specific type, then [see Guide 8 and 9](#).
- If you have a specific type of soft tissue sarcoma, then [see Guide 10](#).
- Alveolar soft part sarcoma (ASPS), atypical lipomatous tumor/well-differentiated liposarcoma (ALT/WDLS), and clear cell sarcomas do not usually respond to chemotherapy.

## Guide 7

### Primary treatment options: Stage 2 and 3 resectable with good function

<b>Stage 2</b>	Surgery
	Surgery followed by radiation therapy
	Radiation therapy before surgery
<b>Stage 3</b>	Surgery followed by <ul style="list-style-type: none"> <li>• Radiation therapy or</li> <li>• Radiation therapy with systemic therapy</li> </ul>
	Radiation therapy before surgery. Consider systemic therapy after surgery.
	Chemoradiation before surgery. Surgery followed by <ul style="list-style-type: none"> <li>• Radiation therapy or</li> <li>• Radiation therapy with systemic therapy</li> </ul>
	Systemic therapy before surgery

**Guide 8****Combination systemic therapies for non-specific subtypes**

Doxorubicin and dacarbazine (called AD)

Doxorubicin, ifosfamide, and mesna (called AIM)

Mesna, doxorubicin, ifosfamide, and dacarbazine (called MAID)

Ifosfamide, epirubicin, and mesna

Gemcitabine and docetaxel

Gemcitabine and vinorelbine (for palliative therapy)

Gemcitabine and dacarbazine

**Guide 9****Single-agent systemic therapies for non-specific subtypes**

Doxorubicin

Ifosfamide

Epirubicin

Gemcitabine

Dacarbazine

Liposomal doxorubicin

Temozolomide (for palliative therapy)

Vinorelbine (for palliative therapy)

Eribulin (for palliative therapy)

Trabectedin (for palliative therapy)

Pazopanib (for palliative therapy)

Regorafenib (for non-adipocytic sarcoma)

Larotrectinib (for *NTRK* gene fusion-positive sarcomas)

Entrectinib (for *NTRK* gene fusion-positive sarcomas)

## Guide 10 Systemic therapies for specific subtypes

<b>Alveolar soft part sarcoma (ASPS)</b>	<ul style="list-style-type: none"> <li>• Sunitinib</li> <li>• Pazopanib</li> <li>• Pembrolizumab</li> </ul>
<b>Angiosarcoma</b>	<ul style="list-style-type: none"> <li>• Paclitaxel</li> <li>• Docetaxel</li> <li>• Vinorelbine (for palliative therapy)</li> <li>• Sorafenib</li> </ul> <ul style="list-style-type: none"> <li>• Sunitinib</li> <li>• Bevacizumab</li> <li>• Other systemic therapies (see Guide 8 and 9)</li> </ul>
<b>Epithelioid sarcoma</b>	<ul style="list-style-type: none"> <li>• Tazemetostat</li> </ul>
<b>Inflammatory myofibroblastic tumor (IMT) with anaplastic lymphoma kinase (ALK) translocation</b>	<ul style="list-style-type: none"> <li>• Crizotinib</li> <li>• Ceritinib</li> </ul>
<b>PEComa, recurrent angiomyolipoma, and lymphangioliomyomatosis</b>	<ul style="list-style-type: none"> <li>• Sirolimus</li> <li>• Everolimus</li> <li>• Temsirolimus</li> </ul>
<b>Solitary fibrous tumor/hemangiopericytoma</b>	<ul style="list-style-type: none"> <li>• Bevacizumab and temozolomide</li> <li>• Sunitinib</li> <li>• Sorafenib</li> <li>• Pazopanib</li> </ul>
<b>Tenosynovial giant cell tumor/pigmented villonodular synovitis</b>	<ul style="list-style-type: none"> <li>• Pexidartinib</li> <li>• Imatinib</li> </ul>
<b>Undifferentiated pleomorphic sarcoma (UPS)</b>	<ul style="list-style-type: none"> <li>• Pembrolizumab</li> </ul>
<b>Well-differentiated/dedifferentiated liposarcoma (WD-DDLS)</b>	<ul style="list-style-type: none"> <li>• Palbociclib</li> </ul>

**Unresectable or resectable with poor function**

For all other tumors that are unresectable or are stage 2 and 3 resectable with poor function, see [Guide 11](#).

Sometimes with stage 2 and 3 tumors, surgery may not be an option because of the location and/or size of the tumor. However, some treatments may shrink the tumor so that surgery is possible.

These treatments include:

- Radiation therapy
- Chemoradiation
- Systemic therapy
- Regional limb therapy

After treatment, your doctors will assess if you are able to have surgery with good results.

Good results include a surgical margin larger than 1 cm, cancer-free margins, and good use of your limb or other body part. This means a

tumor that was unresectable is now resectable. A stage 2 or 3 tumor that is expected to have poor function after surgery might be amputated or receive radiation therapy instead.

**Radiation therapy**

Radiation treatment would be EBRT and the treatment planning should use IMRT, tomography, or protons.

**Chemoradiation**

Treatment that combines chemotherapy with radiation therapy is called chemoradiation.

**Guide 11****Primary treatments: Stage 2 and 3 resectable with poor function, or unresectable**

If resectable with good function, see [Guide 7](#)

- Radiation therapy
- Chemoradiation
- Systemic therapy
- Regional limb therapy
- If amputation, then see follow-up care



If resectable with poor function, then amputation or radiation therapy

If unresectable, then

- Radiation therapy, if not used before
- Systemic therapy
- Palliative surgery
- Observation, if have no symptoms
- Best supportive care

**Regional limb therapy**

Isolated limb infusion/perfusion is another option to treat sarcoma of the limb. It is a type of regional limb therapy. An isolated limb infusion/perfusion is a procedure used to deliver anti-cancer drugs directly into an arm or leg. During surgery, the flow of blood is stopped

for a short period of time while the drugs are injected into the blood of the limb (infusion). This limits the effect of chemotherapy on the rest of the body. This option should only be done at centers or hospitals with experience in regional limb therapy.

**Guide 12****Follow-up testing: Stage 2 and 3**

<b>Recommended</b>	Evaluation for rehabilitation such as occupational therapy and physical therapy
	Medical history and physical exam every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years. Every year afterwards.
	Re-imaging after surgery using MRI with and without contrast (preferred for extremity imaging) or CT with contrast to assess primary tumor and rule out metastatic disease
	Chest imaging using x-ray or CT every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years. Every year afterwards.
	Imaging of the primary site using MRI with or without contrast and/or CT with contrast
	Ultrasound of small lesions near the surface of the skin done by someone experienced in muscle/skeletal disease
<b>Consider</b>	PET/CT may be useful in determining response to neoadjuvant chemotherapy
	CT of abdomen/pelvis for myxoid/round cell liposarcoma, epithelial sarcoma, angiosarcoma, and leiomyosarcoma
	MRI of total spine for myxoid/round cell liposarcoma
	Central nervous system imaging with MRI or CT for alveolar part sarcoma and angiosarcoma
	CT of pelvis for lower-limb well-differentiated liposarcoma

### Observation

If you have no symptoms from the cancer, you may undergo observation. Observation is a period of regular testing for cancer growth so treatment can be started if needed.

### Best supportive care

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

### Amputation

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

### Follow-up care

Follow-up care will begin when treatment is complete. Tests will look for signs of cancer that has returned (recurrence). Follow-up tests can help find cancer early. [See Guide 12.](#)

Follow-up tests include a medical history, physical exam, and imaging tests. You will have a medical history and physical exam every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years, followed by once a year. You should be evaluated for rehabilitation, such as occupational therapy to help with daily life skills or physical therapy to help your body move and function.

If the cancer is likely to return, you may have regular imaging tests of the site where the primary tumor was originally found.

For a list of systemic therapy options, see [Guide 8](#), [9](#), and [10](#).

## Stage 4

In stage 4 sarcoma, the tumor can be any size (T), any grade (G), and there is cancer in nearby lymph nodes (N1). Cancer may or may not have spread to distant parts of the body (M1 or M0). Treatment is based on if the cancer is confined (limited to one organ) or widespread (metastatic).

### Confined cancer

#### Any T, N1, M0, Any G

A stage 4 sarcoma can be any size (T), any grade (G), with cancer in nearby lymph nodes (N1). It has not metastasized to distant sites (M0). The sarcoma is confined to one organ and isn't too big or bulky. This means it appears that the tumor will respond to treatment. In this case, local therapies are used to treat the primary tumor. Local treatments include surgery, radiation therapy, ablation, or embolization. Treatment options are found in [Guide 13](#).

### Metastasectomy

A metastasectomy is different from a mastectomy (surgery to remove the breast). A metastasectomy is surgery to remove one or more metastases. This surgery may be used when the primary tumor can be completely removed and there is only one metastasis. Not all metastases can be removed by surgery. Location, the amount of tissue that must be removed, and your overall health are factors.

A metastasectomy may be done at the same time as surgery to remove the primary tumor. Or, it may be done during a separate operation. The amount of time needed for the surgery and recovery depends on many factors. Some factors include the size and location of the metastases. You may be given systemic therapy before or after this surgery. Stereotactic body radiation therapy (SBRT) may be added.

### Observation

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.

## Guide 13

### Treatment for confined cancer: Stage 4 or recurrence

Treat primary tumor ([see Guide 7](#)) and consider the following:

- Metastasectomy
- Metastasectomy with radiation therapy
- Metastasectomy with pre- or post-surgery systemic therapy (radiation therapy might be added)
- SBRT alone or with systemic therapy
- Ablation
- Embolization
- Observation

## Widespread cancer

### Any T, N1, M1, Any G

A stage 4 sarcoma can be any size (T), any grade (G), with cancer in nearby lymph nodes (N1). It has metastasized to distant sites (M1). These metastases are referred to as disseminated or widespread throughout the body. Treatment is to palliate or to relieve pain and discomfort. Palliative treatment options are found in [Guide 14](#).

### Palliative care

Palliative care does not try to cure the cancer. It is used to relieve pain and discomfort, to reduce symptoms caused by the cancer, and to extend life. Systemic therapy, radiation therapy, and surgery may reduce cancer symptoms by stopping or slowing tumor growth. Other options include ablation and embolization.

### Radiation therapy

Radiation therapy and stereotactic radiation therapy or SBRT are options.

### Observation

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.

### Supportive care

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

## Guide 14

### Palliative care treatment options for metastases: Stage 4 or recurrence

Systemic therapy (see [Guide 8](#), [9](#), and [10](#))

Radiation therapy or SBRT

Surgery

Observation, if have no symptoms

Supportive care

Ablation

Embolization



**Follow-up care**

Follow-up care will begin when treatment is complete. Tests will look for signs of cancer that has returned called recurrence. Follow-up tests can help find cancer early. [See Guide 15.](#)

If needed, you will receive rehabilitation care. This may include occupational or physical therapy.

Tests include a medical history, physical exam, and imaging of the primary tumor site and/or the chest. You may have imaging tests of the primary site right after surgery. If the cancer is likely to return, you may have regular imaging tests of the area where the primary tumor was found before being removed.

**Recurrence**

A recurrence means cancer has returned after a disease-free period of time. It is possible for sarcoma to return after cancer treatment is finished and the sarcoma is in remission. Remission is a period of being free of cancer. If sarcoma does come back, it may return in certain areas. It may be local (in or close to where it started), in one organ, in lymph nodes, or widespread. If it is close to where it started, it is called local recurrence and will be treated as stage 1, 2, or 3.

If disease is metastatic, then treatment is based on if metastases are:

- Confined to one organ
- Widespread
- Isolated to a specific region or found in lymph nodes

**Guide 15****Follow-up testing: Stage 4**

Evaluation for rehabilitation such as occupational therapy and physical therapy.

Medical history and physical exam every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years. Every year afterwards.

Chest imaging and other known metastatic sites every 2 to 6 months for 2 to 3 years, then every 6 months for the next 2 years. Every year afterwards in those with no evidence of disease. X-ray or CT is preferred.

Consider imaging of the primary site using MRI with or without contrast and/or CT with contrast.

Consider ultrasound of small lesions near the surface of the skin done by someone experienced in muscle/skeletal disease.

Treatment options are guided by where the cancer has returned. For example, a local recurrence may be treated like a newly diagnosed soft tissue sarcoma.

- Confined cancer recurrence will be treated as in [Guide 13](#).
- Widespread cancer recurrence will be treated as in [Guide 14](#).
- Cancer that is isolated to a specific region or found in lymph nodes will be treated as in [Guide 16](#).

### Isolated regional disease or in lymph nodes

In isolated regional disease, a soft tissue sarcoma has returned near the site of the first (primary) tumor. Cancer might be found in lymph nodes close to where the cancer started, but it has not spread to distant sites in the body. Treatment options are found in [Guide 16](#).

### Regional lymph node dissection

A lymph node dissection is an operation to remove lymph nodes from your body. It might be followed by radiation therapy with or without systemic therapy.

### Metastasectomy

A metastasectomy is surgery to remove one or more metastases. Systemic therapy might be given before surgery (neoadjuvant) or after surgery (adjuvant). Radiation therapy might be added.

### SBRT

Stereotactic radiation therapy (SBRT) is an option. SBRT uses high-energy radiation beams to treat cancers.

### Isolated limb infusion/perfusion

An isolated limb infusion/perfusion is a procedure used to deliver anti-cancer drugs directly into an arm or leg. During surgery, the flow of blood is stopped for a short period of time while the drugs are injected into the blood of the limb (infusion). This procedure should be done at hospitals or centers with experience using this treatment.

## Guide 16

### Recurrence treatment options: Isolated disease or found in lymph nodes

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Regional lymph node dissection (surgery to remove lymph node)

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Regional lymph node dissection with radiation therapy (systemic therapy might be added)

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Metastasectomy

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Metastasectomy with pre- or post-surgery systemic therapy (radiation therapy might be added)

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SBRT

---

Isolated limb perfusion/infusion with surgery

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## Review

- Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating your type of soft tissue sarcoma.
- Most stage 1 sarcomas in the limbs, outer torso, head, or neck can be treated with surgery.
- Stage 2 and 3 sarcomas may or may not be treated with surgery.
- Observation is a period of regular testing for cancer growth so treatment can be started if needed.
- Treatment for stage 4 sarcomas depends on if the metastases are in one organ or widespread.
- A metastasectomy is surgery to remove one or more metastases.
- Recurrence means cancer has returned after a disease-free a period of time.
- Treatment options for recurrences are based on the extent or amount of cancer.
- A lymph node dissection is an operation to remove lymph nodes from your body.
- An isolated limb infusion/perfusion is a procedure used to deliver anti-cancer drugs directly into an arm or leg.

## Order of treatments

Most people with soft tissue sarcoma will receive more than one type of treatment. Below is an overview of the order of treatments and what they do.



### **Neoadjuvant (before) treatment**

is given to shrink the tumor before primary treatment (surgery).



**Primary treatment** is the main treatment given to rid the body of cancer. Surgery is usually the main treatment for soft tissue sarcoma.



**Adjuvant (after) treatment** is given after primary treatment to rid the body of any cancerous or abnormal cells left behind from surgery. It is also used when the risk of cancer returning (recurrence) is felt to be high.

# 5

## Sarcomas inside the torso

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51 Tests

---

52 Stages

---

54 Resectable

---

56 Unresectable or stage 4

---

57 Review



Sarcomas found in organs inside the chest, abdomen, or pelvis are referred to as the inner trunk (torso). Surgery is usually the main treatment. Together, you and your doctor will choose a treatment plan that is best for you.

## Tests

You will have a variety of tests based on the suspected type of soft tissue sarcoma and to rule out desmoid tumors and other lesions. Doctors will take a medical history, complete a physical exam of your body, and order imaging tests. A biopsy should be done to confirm cancer and determine the specific type of

soft tissue sarcoma. A core needle biopsy is preferred. For recommended tests, see [Guide 17](#).

Testing is needed to diagnose soft tissue sarcoma. Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating your type of soft tissue sarcoma.

If you have a personal or family history of a rare hereditary cancer syndrome called Li-Fraumeni syndrome, NCCN experts suggest a genetics assessment. This assessment includes meeting with a genetic counselor who will talk with you about the results.

### Guide 17 Tests for sarcomas in the inner torso

<b>Needed</b>	Before starting therapy, all patients should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma.
	Medical history and physical exam
	CT of chest/abdomen/pelvis. MRI of abdomen/pelvis might also be done.
	Image-guided core needle biopsy should be performed if preoperative (neoadjuvant) therapy is being given or for suspicion of malignancy other than sarcoma. Pre-surgery biopsy is not required for well-differentiated liposarcoma.
	Chest imaging such as x-ray or CT without contrast (preferred)
<b>Useful</b>	Consider more genetic testing in those with a personal or family history of Li-Fraumeni syndrome.
	Other testing for those with neurofibromatosis type 1

## Stages

### Visceral organs of the abdomen and chest

TNM staging and grade are used to diagnose and treat soft tissue sarcomas found in the visceral organs of the abdomen and chest. Visceral organs include any soft organ, but not lymph nodes. Since there are many possible organs that could have a sarcoma, traditional staging is difficult. Therefore, sarcomas found in these areas are not divided into stages. TNM scores and tumor grades for soft tissue sarcomas of the abdomen and chest are described next.

#### T = Tumor

The primary tumor size is measured in centimeters (cm). A large pea is 1 cm. A golf ball is 4 cm. T4 tumors are multifocal. In multifocal cancer, there is more than one tumor, but none are considered primary. In metastatic cancer, there is one primary tumor with smaller tumors elsewhere.

- **T1** Organ confined
- **T2** Tumor extends beyond the organ
- **T3** Tumor invades another organ
- **T4** There are tumors in 2 or more sites

#### N = Node

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are those located near the tumor.

- **N0** means no regional lymph node involvement or unknown
- **N1** means tumor is found in nearby lymph nodes

#### M = Metastatic

Cancer that has spread to distant parts of the body is shown as M1. The most common site for metastasis is the liver or lung.

- **M0** means no distant metastasis
- **M1** means distant metastasis is found

#### G = Grade

Grade describes how abnormal the tumor cells look under a microscope and how quickly these cells are likely to grow and spread. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. G3 is the highest grade.

- **GX** means grade cannot be assessed
- **G1** is a score of 2 or 3
- **G2** is a score of 4 or 5
- **G3** is a score of 6, 7, or 8

### Retroperitoneum

The peritoneum is the lining that surrounds the abdominal organs. The retroperitoneum is the area behind and outside of the peritoneum. It is a complex part of the body that includes organs such as major blood vessels, adrenal glands, kidneys, pancreas, and bladder. Liposarcoma and leiomyosarcoma are the most common types of soft tissue sarcoma found in the retroperitoneum.

Tumors in the retroperitoneum tend to grow very large before causing symptoms. These tumors tend to push against or invade nearby structures such as veins and arteries. The size of the tumor and location to major blood vessels makes surgery difficult. Staging is based imaging and biopsy. A biopsy is needed to confirm soft tissue sarcoma in the retroperitoneum. For stages, [see Guide 18](#).

**T = Tumor**

The primary tumor size is measured in centimeters (cm). A golf ball is 4 cm. A baseball is 7 cm. A grapefruit is 15 cm.

- **T1** Tumor is 5 cm or less
- **T2** Tumor is 5.1 cm to 10 cm
- **T3** Tumor is 10.1 cm to 15 cm
- **T4** Tumor is more than 15 cm

**N = Node**

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are those located near the tumor. Metastasis in lymph nodes is unusual.

- **N0** means no regional lymph node metastasis or unknown
- **N1** means regional lymph node metastasis is found

**Guide 18****Cancer stages for soft tissue tumors in the retroperitoneum**

<b>Stage 1</b>	<b>Stage 1A</b>	• T1, N0, M0, G1 or GX
	<b>Stage 1B</b>	• T2, N0, M0, G1 or GX • T3, N0, M0, G1 or GX • T4, N0, M0, G1 or GX
<b>Stage 2</b>	• T1, N0, M0, G2 or G3	
<b>Stage 3</b>	<b>Stage 3A</b>	• T2, N0, M0, G2 or G3
	<b>Stage 3B</b>	• T3, N0, M0, G2 or G3 • T4, N0, M0, G2 or G3
<b>Stage 4</b>	• Any T, N1, M0, Any G • Any T, Any N, M1, Any G (metastatic)	

**M = Metastatic**

Cancer that has spread to distant parts of the body is shown as M1. The most common site for metastasis is the lung.

- **M0** means no distant metastasis
- **M1** means distant metastasis is found

**G = Grade**

Grade describes how abnormal the tumor cells look under a microscope and how quickly these cells are to grow and spread. Higher-grade sarcomas tend to grow and spread faster than lower-grade sarcomas. G3 is the highest grade.

- **GX** means grade cannot be assessed
- **G1** is a score of 2 or 3
- **G2** is a score of 4 or 5
- **G3** is a score of 6, 7, or 8

**Resectable**

A soft tissue sarcoma that can be removed with surgery is called resectable. This is usually the main treatment. A biopsy may or may not be done before surgery. If you will have treatment before surgery (neoadjuvant), then an image-guided core needle biopsy is needed.

**Before surgery**

Neoadjuvant therapy might include radiation therapy or systemic therapy. Surgery is the primary treatment. If neoadjuvant therapy is an option, then proof of subtype is needed from a biopsy before treatment can begin.

**During surgery**

During surgery, intraoperative radiation therapy (IORT) might be given. A sample of the tumor and surgical margin may be sent to a pathologist for review. An experienced sarcoma pathologist should review any biopsy and tissue samples.

Complete resection with a negative surgical margin (R0) can be difficult to achieve. One reason is the tumor is near vital organs, veins, arteries, and other tissues. For a list of systemic therapy options, [see Guide 8, 9, and 10](#).

**After surgery**

After surgery, your doctors will assess if you need further treatment such as adjuvant treatment. Adjuvant treatment may kill any remaining cancer cells and help stop the cancer from returning. [See Guide 19](#).

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

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



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

### Follow-up care

You will have follow-up tests to check if the cancer has returned. Follow-up tests include a physical exam and CT of the abdomen/pelvis or MRI every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years, followed by every year afterwards. If the cancer is likely to spread to your lungs, you may get imaging tests of the chest, such as an x-ray or CT (preferred). [See Guide 19.](#)

For a list of systemic therapy options, see [Guide 8, 9, and 10.](#)

## Guide 19

### After surgery treatment based on surgical margin results

<b>No disease remains (R0)</b>	Radiation therapy in certain cases	Follow-up testing <ul style="list-style-type: none"> <li>• Physical exam with imaging every 3 to 6 months for 2 to 3 years, then every 6 months for the next 2 years. Every year afterwards.</li> <li>• Imaging includes CT of abdomen/pelvis or MRI</li> <li>• Chest imaging: X-ray or CT (preferred)</li> </ul>
<b>Some disease remains (R1)</b>	Radiation therapy in certain cases	
	In certain cases, consider brachytherapy boost if radiation given before surgery	
<b>Disease remains (R2)</b>	Consider re-section for low-grade disease or well-differentiated liposarcoma	
	See <a href="#">Guide 20</a>	

## Unresectable or stage 4

A tumor that cannot be removed with surgery is called unresectable. These include tumors that involve vital structures or tumors whose removal could cause complications or death. Overall health also plays a role. One must be medically fit to tolerate major abdominal surgery.

Stage 4 tumors can also be unresectable. Stage 4 tumors can be any size (T), any grade (G), with cancer in nearby lymph nodes (N1). It has metastasized to distant sites (M1).

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

Sometimes, an unresectable tumor can become resectable. An attempt might be made to shrink the tumor using systemic therapy, chemoradiation, or radiation therapy. Imaging tests will be used to see if the tumor has shrunk and if surgery is now possible.

### Palliative care

Palliative care does not try to cure the cancer; it aims to relieve your discomfort. Systemic therapy, radiation therapy, and surgery may reduce cancer symptoms by stopping or slowing tumor growth. Other options include ablation and embolization. For palliative care options, [see Guide 20](#).

### Follow-up care

You will have follow-up tests to see how the tumor is responding to treatment.

These might include:

- CT of chest, abdomen, and pelvis
- Chest CT without contrast
- MRI of abdomen and pelvis

## Guide 20

### Palliative care options: Unresectable, stage 4, or disease progression

Systemic therapy

Radiation therapy

Surgery for symptom control

Supportive care

Observation, if not having symptoms

## Review

- ▶ Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating your type of soft tissue sarcoma.
- ▶ A soft tissue sarcoma that can be removed with surgery is called resectable. This is usually the main treatment for soft tissue sarcomas in the inner torso.
- ▶ A tumor that cannot be removed with surgery is called unresectable.
- ▶ Palliative care does not try to cure the cancer; it aims to relieve your discomfort.
- ▶ You will have tests to see how the tumor is responding to treatment. Follow-up tests might include a physical exam and imaging of your chest, abdomen, and pelvis.

Get to know your  
care team and let  
them get to  
know you.

# 6

## Gastrointestinal stromal tumors

59	Overview
60	Tests
62	Stages
64	Stomach <b>GIST</b> of less than 2 cm
65	All other GISTs
68	Disease progression
70	Review



Gastrointestinal stromal tumors (GISTs) are soft and fragile tumors that usually start in the walls of the gastrointestinal tract. The gastrointestinal tract includes the esophagus, stomach, small intestine, colon, and rectum. Together, you and your doctor will choose a treatment plan that is best for you.

## Overview

Gastrointestinal stromal tumor (GIST) is the most common sarcoma and the most common soft tissue sarcoma of the gastrointestinal (GI) tract. The gastrointestinal tract includes the esophagus, stomach, small intestine, colon, and rectum. GISTs are most commonly found in the stomach and small intestine.

The stomach is an organ that is part of the digestive system. It helps digest food. The small intestine connects the stomach with the large intestine. It is about 20 feet long and is folded inside the abdomen (belly).

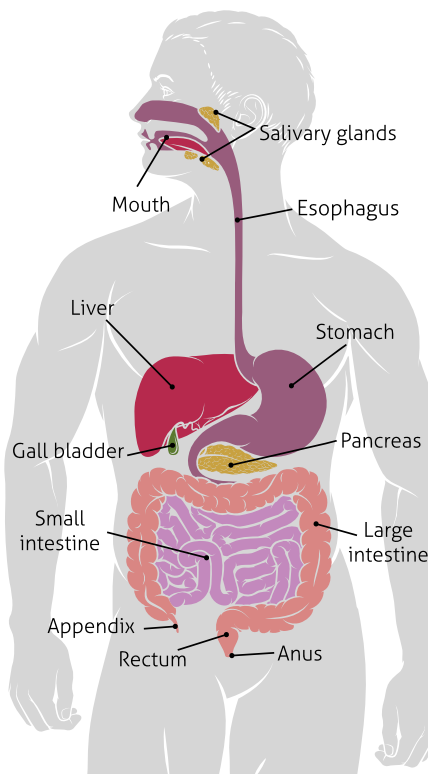
There are 3 parts of the small intestine that help to further digest food coming from the stomach:

- **Duodenum** –first part
- **Jejunum** – middle part
- **Ileum** – last part

A GIST is a specific type of tumor. These tumors can be very small and grow slowly or large and grow quickly. Tumors that are smaller than 2 cm usually grow slowly and are usually benign (not cancer). They are usually not aggressive. However, some will grow fast and may become a problem.

### Gastrointestinal stromal tumor (GIST)

**GISTs are found in the stomach or surrounding organs and tissues, such as the small intestine.**



## Tests

Testing is needed to diagnose GIST. You will have a biopsy and imaging tests. Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating GIST.

GIST is divided into those found in the stomach that are less than 2 cm and all others. Testing for GIST can be found in [Guide 21](#).

### Biopsy

A biopsy is needed to confirm GIST. The pathology report should include location, size, and accurate measurement of mitotic rate in the fastest growing part of tumor.

An endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) is the preferred type of biopsy for a very small GIST (less than 2 cm) in the stomach. An EUS-FNA uses a thin, tube shaped tool called an endoscope that is inserted through the mouth and into the

### Guide 21 Tests for GIST

Before starting therapy, all patients should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma

<b>Very small GIST of less than 2 cm</b>	CT of abdomen/pelvis and/or MRI
	Endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA)
<b>All other GISTs</b>	CT of abdomen/pelvis and/or MRI of abdomen/pelvis
	Consider chest imaging using x-ray or CT
	Biopsy to confirm GIST
	Testing for mutations in <i>KIT</i> and <i>PDGFRA</i> is strongly recommended
	If tumor is lacking <i>KIT</i> and <i>PDGFRA</i> , then recommend testing for SDHB using immunohistochemistry (IHC)
	Testing for mutations in the <i>SDH</i> genes should be considered for those with wild-type GIST (lacking <i>KIT</i> and <i>PDGFRA</i> mutations) who are SDH-deficient
Genotyping should be performed when medical therapy is planned	

body to take pictures. One end of the scope has a small light and camera lens to see inside your body. The other end has a probe, which bounces sound waves off of your stomach and other organs. The image is sent to a television monitor and is used to guide the biopsy. A sample of the GIST is removed with a needle.

### Genetic tests

Your doctor may also suggest tests for gene mutations. Genes tell cells what to become and what to do. In a process called mutation something goes wrong in the genetic code. This can cause cells to grow and divide out of control.

Genetic testing looks for the presence or absence of certain gene mutations. Gene mutations in GIST may include *KIT*, *PDGFRA*, and *SDH*. Test results will help your treatment team learn more about the tumor and make a treatment plan.

### *KIT* gene mutations

Mutations in the *KIT* gene are the most common genetic changes found in GIST. In most cases, *KIT* gene mutations are acquired or happen during a person's lifetime. This mutation causes cells to grow. Certain targeted therapies are used to treat GIST with *KIT*.

### *PDGFRA* gene mutations

Mutations in the platelet-derived growth factor receptor alpha (*PDGFRA*) gene are often found in GIST of the stomach. In most cases, *PDGFRA* gene mutations are acquired or happen during a person's lifetime. This mutation causes cells to grow. Certain targeted therapies such as avapritinib are used to treat GIST with *PDGFRA*.

### *SDH* gene mutations

Succinate dehydrogenase (SDH) is a protein within cells that converts energy. There are 4 parts to SDH: SDHA, SDHB, SDHC, and SDHD. If you are diagnosed with a gastrointestinal stromal tumor (GIST) that doesn't have *KIT* or *PDGFRA* gene mutation, then additional genetic testing will be considered to look for mutations in *SDH* genes or to see if you are lacking SDHD.

GISTs with *SDH* mutation arise in the stomach in younger individuals, frequently metastasize, may involve lymph nodes, and usually grow slowly. They are usually resistant to imatinib.

### Wild-type GIST

Wild-type GIST do not have or lack *KIT* or *PDGFRA* mutations. Testing for *SDH* mutations should be considered for wild-type GIST.

### Immunohistochemistry

Immunohistochemistry (said immuno-histo-chemistry or IHC) is a special staining process that involves adding a chemical marker to cells. The cells are then studied using a microscope.

### Genotyping

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

## Stages

The TNM staging system is used for GIST. Staging is based on testing. Staging does not include mutations, but does take into account how fast the cells make copies of themselves (mitotic rate). For stages, [see Guide 22 and 23](#).

### T = Tumor

The primary tumor size is measured in centimeters (cm). A golf ball is 4 cm. A baseball is 7 cm. A grapefruit is 15 cm.

- **T0** No evidence of a tumor
- **T1** Tumor is 2 cm or less
- **T2** Tumor is 2.1 cm to 5 cm
- **T3** Tumor is 5.1 cm to 10 cm
- **T4** Tumor is more than 10 cm

### N = Node

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are those located near the tumor. Lymph node metastasis in GIST are rare, except in SDH-deficient GISTs, which tend to be less aggressive than other GISTs.

- **N0** means no regional lymph node metastasis or unknown
- **N1** means regional lymph node metastasis is found

### M = Metastatic

Cancer that has spread to distant parts of the body is shown as M1. The most common site for metastasis is the liver or the peritoneum (the membrane that lines the abdomen).

- **M0** means no distant metastasis
- **M1** means distant metastasis is found

### Mitotic rate

Mitotic rate is the speed at which cancer cells make copies of themselves. A low mitotic rate is slower than a high mitotic rate.



## Guide 22

## Cancer stages for GIST in stomach

Stage 1	Stage 1A	• T1 or T2, N0, M0, low mitotic rate
	Stage 1B	• T3, N0, M0, low mitotic rate
Stage 2		<ul style="list-style-type: none"> <li>• T1, N0, M0, high mitotic rate</li> <li>• T2, N0, M0, high mitotic rate</li> <li>• T4, N0, M0, low mitotic rate</li> </ul>
Stage 3	Stage 3A	• T3, N0, M0, high mitotic rate
	Stage 3B	• T4, N0, M0, high mitotic rate
Stage 4		<ul style="list-style-type: none"> <li>• Any T, N1, M0, Any rate</li> <li>• Any T, Any N, M1, Any rate (metastatic)</li> </ul>

## Guide 23

## Cancer stages for GIST in small intestine

Stage 1		• T1 or T2, N0, M0, low mitotic rate
Stage 2		• T3, N0, M0, low mitotic rate
Stage 3	Stage 3A	<ul style="list-style-type: none"> <li>• T1, N0, M0, high mitotic rate</li> <li>• T4, N0, M0, low mitotic rate</li> </ul>
	Stage 3B	<ul style="list-style-type: none"> <li>• T2, N0, M0, high mitotic rate</li> <li>• T3, N0, M0, high mitotic rate</li> <li>• T4, N0, M0, high mitotic rate</li> </ul>
Stage 4		<ul style="list-style-type: none"> <li>• Any T, N1, M0, Any rate</li> <li>• Any T, Any N, M1, Any rate (metastatic)</li> </ul>

## Stomach GIST of less than 2 cm

A stomach GIST of less than 2 cm is treated differently than other GISTs. Most often, these small tumors are removed with surgery. They are almost always benign (not cancer).

### Biopsy

An endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) is the preferred type of biopsy for a very small GIST (less than 2 cm) in the stomach. It is need to confirm the diagnosis of the GIST. Imaging will also be done.

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

Surgery to remove the GIST is called surgical resection. You might have imatinib before surgery to shrink the tumor.

Tumors that aren't high risk don't need treatment. Instead, the tumor might be monitored with endoscopic or imaging tests. This option has risks. Discuss with your doctor if surveillance is the best option for you.

### After surgery

After surgery, most people take imatinib. However, since not every GIST responds to imatinib in the same way, you might take a different systemic therapy. Treatment options can be found in [Guide 24](#).

## Guide 24

### Post-surgery treatment based on surgical margin results

<b>GIST removed with surgery (R0)</b>	<ul style="list-style-type: none"> <li>• Imatinib for those at high risk of recurrence</li> <li>• Observation for low-risk disease</li> </ul>	
<b>Imatinib first, then GIST removed with surgery (R0)</b>	<ul style="list-style-type: none"> <li>• Consider continuing imatinib</li> </ul>	
<b>Disease remains (R2)</b>	If had imatinib before surgery, then continue on imatinib and consider more surgery	Continue imatinib or avapritinib until disease progression
	If didn't have imatinib before surgery, then start imatinib	
	If <i>PDGFRA</i> mutation, then start avapritinib and consider surgery	
<b>Metastatic disease</b>	See <a href="#">Guide 27</a>	

**Follow-up care**

You will be monitored closely. You may get CT scans of your abdomen and pelvis. It is suggested the CT scans occur every 3 to 6 months for 3 to 5 years. If test results are normal for 3 to 5 years, you will have a CT scan every year. A PET/CT might be done if CT results are unclear. An MRI is also possible.

See Guide 25.

**All other GISTs**

GISTs are most common in the stomach or small intestine. Very small GISTs found outside the stomach can be aggressive. These tumors can grow quickly, regrow after treatment, spread to nearby lymph nodes, or metastasize. However, this is not always the case. This is why it is important that test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating GISTs.

In general:

- GISTs of the small intestine tend to be more aggressive than those found in the stomach.
- GISTs of the colon tend to be aggressive and tumors less than 2 cm can metastasize.

**Biopsy**

A biopsy is needed to confirm the diagnosis of the GIST. Your doctors will assess the tumor based on imaging tests and the pathology report. The pathology report will include the size, location, and mitotic rate of the GIST. For GISTs that have complex or unusual histology, referral to a center that is experienced in sarcoma diagnosis is recommended.

Depending on the size of the tumor and its location, surgery can be very difficult. Surgery will likely affect nearby organs and tissues. There is risk. Treatment is based on if the tumor is resectable, unresectable, or metastatic.

**Guide 25****Follow-up testing: GIST**

Medical history and physical exam every 3 to 6 months.

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For completely resected GIST, CT of abdomen/pelvis every 3 to 6 months for 3 to 5 years, then every year afterward. Testing might be less often for low-risk or very small tumors of less than 2 cm.

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For incompletely resected GIST or if metastasis found during surgery, then CT of abdomen/pelvis every 3 to 6 months.

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For disease progression, CT, MRI, or PET/CT may be done.

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After treatment for disease progression, check therapy response with CT of abdomen/pelvis, or MRI. PET/CT might be done if CT results are unclear.

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### Resectable

Surgery to remove or resect the primary tumor can be your first treatment if it does not put you at unreasonable risk for complications. Surgery to remove a GIST requires a great deal of skill. GISTs are fragile and easily crumble. In addition, blood vessels might have to be removed or pieces cut out and sewn back together. Parts of organs such as your stomach might have to be removed and sewn back together. All of the tumor must be removed in order to achieve a negative margin resection (R0). This might not be possible. You might have imatinib before surgery to shrink the GIST and help achieve R0.

Your team will gather as much information as possible before surgery. Often, it is very hard to know until surgery how much cancer there is and if there is cancer in any veins, arteries, and other organs. Ask your surgeon what might be removed during surgery and what this means in terms of healing and recovery time.

### Adjuvant therapy (treatment after surgery)

After surgery, you may receive more treatment. This is called adjuvant therapy. Adjuvant therapy is usually imatinib, but it could be sunitinib. Imatinib can cause life-threatening side effects and not be the right option for you. Because those with GISTs respond differently to imatinib, you should have mutation testing to look for *KIT* and *PDGFRA* gene mutations. Avapritinib will be used in those with *PDGFRA* exon 18 mutation, including *PDGFRA* D842V mutations.

If all the cancer appears to be removed, you could start imatinib if there's a high or medium risk for the cancer returning. If your surgeon was unable to remove all of the cancer, you

may start imatinib and consider a second surgery.

Systemic therapies for GIST can be found in [Guide 26](#).

### Follow-up care after surgery

It is important to continue taking your systemic therapy after surgery. You will have follow-up tests after surgery.

If all the tumor was removed, testing will see if the tumor has returned. Follow-up tests can help find cancer early. To find cancer early, get a medical history and physical exam every 3 to 6 months for 5 years. If results are normal for 5 years, then these tests may be done every year. In addition, get a CT of your abdomen and pelvis. If results are normal during this time, the follow-up tests are done once a year.

If not all the GIST was removed, testing can see if cancer remains or is growing. Tests should include medical history, physical exam, and imaging tests of your abdomen and pelvis every 3 to 6 months. If the cancer is growing, see treatment for disease progression.

## Guide 26 Systemic therapies for GIST

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Imatinib

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Sunitinib

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Avapritinib

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Regorafenib

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**Unresectable, recurrent, or metastatic**

Imatinib is used for GIST that can't be removed with surgery (unresectable), returned after surgery (recurrence), or has spread far (metastasized). It is very important that you don't stop taking imatinib once you've started. GIST will worsen if you stop taking your medicine or if you miss doses. If you have GIST with *PDGFRA* exon 18 mutation, including *PDGFRA* D842V mutations, then you will take avapritinib (and not imatinib).

You will have imaging tests to show how the GIST is responding to treatment. If the GIST is stable or shrinking, then surgery might be an option. However, if you were taking imatinib before, then you will continue taking it after surgery.

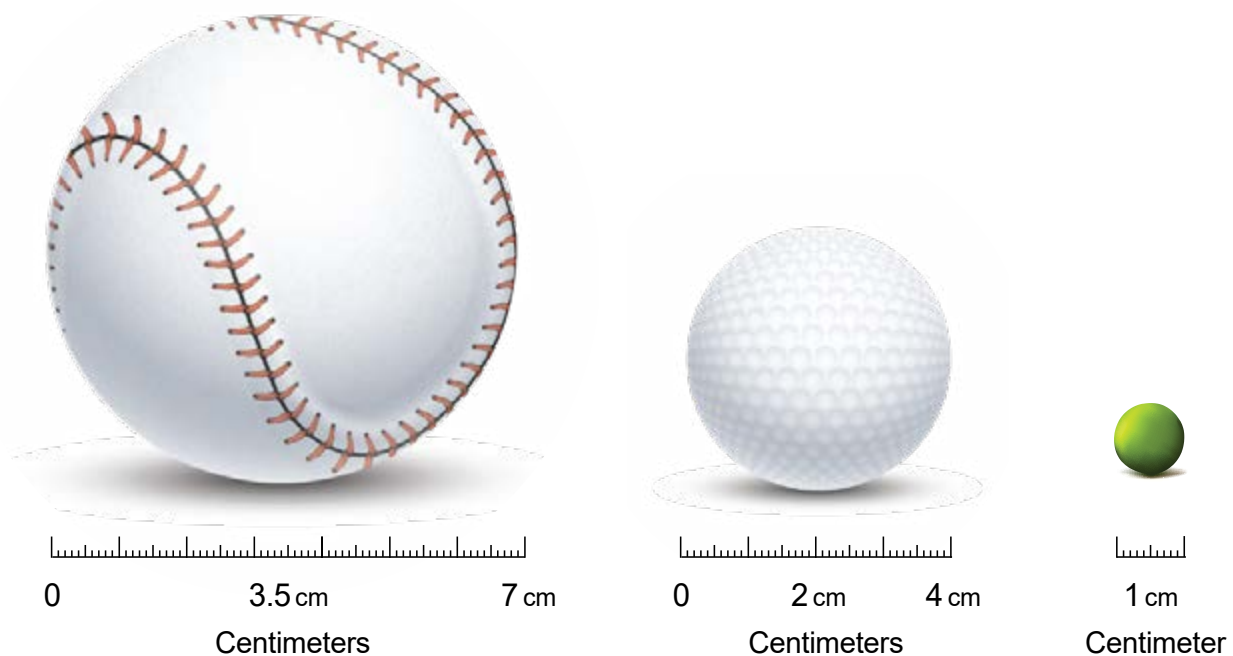
**Metastatic**

Imatinib is the main therapy for metastatic GIST. However, if you have *PDGFRA* exon 18 mutation, including *PDGFRA* D842V mutations, then you will take avapritinib (and not imatinib). Surgery may be possible if tumors shrink or stop growing in response to imatinib. Imatinib can be stopped right before surgery and restarted as soon as a person can tolerate oral medicine.

Tumors can become resistant to imatinib due to changes in the tumor. Sometimes, people cannot tolerate imatinib. In either case, sunitinib will be used. If disease progresses while on imatinib or sunitinib, then regorafenib will be used.

**GISTs are measured in centimeters**

A baseball is 7 cm, a golf ball is 4 cm, and a large pea is 1 cm.



## Disease progression

Some GISTs grow while taking drug treatment. This is called disease progression. Treatment options are based on whether the disease is limited or widespread growth. Treatment options after progression are found in [Guide 27](#).

You will have imaging tests such as a CT or MRI. A PET scan may be done if the other results are unclear. It is recommended that you ask for a referral to a sarcoma speciality center.

### Limited

For limited growth, there are a few different options.

1. Keep taking imatinib or avapritinib and consider the following local treatments:
  - Resection
  - Radiofrequency ablation, embolization, or chemoradiation
  - Palliative radiation therapy
2. Increase the dose of imatinib or change to sunitinib. Sunitinib may be given if the

### Guide 27 Disease progression

<b>Limited</b>	Continue with the same dose of imatinib or avapritinib	If disease progresses despite imatinib, sunitinib, or regorafenib, then consider: <ul style="list-style-type: none"> <li>• Clinical trial</li> <li>• Systemic therapy (<a href="#">see Guide 28</a>)</li> <li>• Best supportive care</li> </ul>
	If GIST grows on avapritinib, then a clinical trial is recommended	
	If GIST grows on imatinib, consider the following: <ul style="list-style-type: none"> <li>• Resection</li> <li>• Radiofrequency ablation, embolization, or chemoembolization</li> <li>• Palliative radiation therapy</li> <li>• Increase dose of imatinib or change to sunitinib</li> </ul>	
	Imaging to check for response to therapy	
<b>Widespread</b>	For performance status (PS) 0, 1, or 2 with disease progression on imatinib, then: <ul style="list-style-type: none"> <li>• Increase dose of imatinib or</li> <li>• Change to sunitinib. If disease progresses on sunitinib, then regorafenib</li> <li>• Imaging to check for response to therapy</li> </ul>	

tumor does not respond to imatinib or the side effects of imatinib are too intense.

Surgery (resection) might be possible. If you are taking imatinib, it can be stopped right before surgery and restarted soon after surgery.

Imaging to check for treatment response might include a CT or MRI of the abdomen and pelvis. A PET scan may be used if the CT or MRI results are unclear.

### Widespread

For widespread tumor growth, options are to increase the dose of imatinib or start taking sunitinib. This decision is based on your performance status (PS). PS is a rating based on your overall health, cancer symptoms, and the ability to do daily activities. Good PS is usually PS 0 or PS 1.

If your PS is 0, 1, or 2 and disease progresses while on imatinib, then:

- Dose of imatinib might be increased or
- Drug might be changed to sunitinib. If disease progresses on sunitinib, then regorafenib will be tried.

Imaging (CT or MRI) will be used to check how treatment is working. A PET/CT may be used if CT or MRI results are unclear.

### Disease progresses on systemic therapy

When GIST spreads or continues to grow despite taking imatinib, sunitinib, or regorafenib, there are 3 treatment options:

- Clinical trial
- Systemic therapy, [see Guide 28](#)
- Best supportive care

### Best supportive care

Best supportive care is used with other treatments to improve quality of life. Best supportive care might include systemic therapies as found in [Guide 28](#). Imatinib, sunitinib, or regorafenib might be reintroduced to help manage symptoms. A clinical trial is the best option for GIST that progresses on avapritinib.

## Guide 28 Systemic therapies for disease progression in GIST

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Sorafenib

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Nilotinib

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Dasatinib (for D842V mutations)

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Pazopanib

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Everolimus with a TKI (imatinib, sunitinib, or regorafenib)

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## Review

- Gastrointestinal stromal tumors (GISTs) are soft and fragile tumors that usually start in the gastrointestinal tract. The gastrointestinal tract includes the esophagus, stomach, small intestine, colon, and rectum.
- Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating GISTs.
- Small stomach GISTs that likely won't grow fast don't need treatment. Instead, these tumors may be watched. This is called observation.
- Small stomach GISTs that likely will grow fast can be removed with surgery. A GIST that can be removed with surgery is called resectable.
- All other GISTs first may be treated with surgery or imatinib.
- If a GIST grows while you're taking imatinib, the dose may be increased or you may be switched to sunitinib. If disease progresses on sunitinib, then regorafenib will be tried.
- You will have tests to see how the tumor is responding to treatment. Tests include a medical history, physical exam, and imaging.
- A clinical trial is the best option for GIST that progresses on avapritinib.
- Best supportive care is used with other treatments to improve quality of life.



### Finding a clinical trial

- Search the National Institutes of Health (NIH) database for clinical trials. It includes publicly and privately funded clinical trials, who to contact, and how to enroll. Look for an open clinical trial for your specific type of cancer. Go to [ClinicalTrials.gov](https://clinicaltrials.gov).
- The National Cancer Institute's Cancer Information Service (CIS) provides up-to-date information on clinical trials. You can call, e-mail, or chat live. Call 1.800.4.CANCER (800.422.6237) or go to [cancer.gov](https://cancer.gov).



# 7

## Desmoid tumors

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72 Overview

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73 Resectable

---

75 Unresectable

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76 Review



Desmoid tumors are dense tumors that do not metastasize. However, since these tumors are aggressive and often recur after treatment, they are treated like cancer. Together, you and your doctor will choose a treatment plan that is best for you.

## Overview

Desmoid tumors are locally aggressive tumors. They tend to return after treatment, often in the same place. This is referred to as locally invasive. Treatment planning considers the type of tumor, whether other types of sarcoma are found, and the tumor location(s). Observation or systemic therapy are the main treatments for desmoid tumors. Desmoid tumors are also known as aggressive fibromatosis or desmoid-type fibromatosis.

## Tests

Testing is needed to diagnose (confirm) a desmoid tumor. Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating desmoid tumors.

Tests for desmoid tumors can be found in [Guide 29](#).

Desmoid tumors often occur for unknown reasons. However, some people have syndromes that increase their chances of getting these tumors. Gardner's syndrome and familial adenomatous polyposis (FAP) are such syndromes. Both are rare disorders that increase the chances for cancer and desmoid tumors. Your doctors will assess if you likely have either of these syndromes.

## Guide 29

### Tests for desmoid tumors

Before starting therapy, all patients should be evaluated and managed by a multidisciplinary team with expertise and experience in sarcoma

Medical history and physical exam

Evaluation for Gardner's syndrome or FAP

Imaging of primary site with CT or MRI, as needed

Biopsy

## Resectable

### Treatment options

A tumor that can be removed with surgery is called resectable. Even though a tumor is resectable, surgery might not be the first treatment option. For treatment options, [see Guide 30](#).

There are 3 treatment options for tumors that are resectable:

- Observation
- Surgery
- Radiation therapy and/or systemic therapy

### Guide 30 Primary treatment: Resectable desmoid tumors

<b>Observation</b>	<ul style="list-style-type: none"> <li>• If tumor is stable, then continue to monitor with imaging, physical exam, and health history</li> <li>• If tumor grows or spreads, then see other options in this guide</li> </ul>	
<b>Surgery (R0)</b>	<ul style="list-style-type: none"> <li>• Observation</li> </ul>	<p>Follow-up care</p> <ul style="list-style-type: none"> <li>• Monitor for symptoms</li> <li>• Evaluate for rehabilitation</li> <li>• Physical exam with medical history and imaging every 3 to 6 months for 2 to 3 years, then every 6 to 12 months thereafter</li> </ul> <p>If disease progresses or returns, then consider:</p> <ul style="list-style-type: none"> <li>• Systemic therapy</li> <li>• Resection</li> <li>• Resection with radiation therapy</li> <li>• Radiation therapy alone</li> <li>• Radiation is not an option if had before</li> </ul>
<b>Surgery (R1)</b>	<ul style="list-style-type: none"> <li>• Observation</li> <li>• Consider re-resection</li> <li>• Adjuvant radiation therapy</li> </ul>	
<b>Surgery (R2)</b>	<ul style="list-style-type: none"> <li>• Radiation therapy</li> <li>• Systemic therapy</li> <li>• Radical surgery if other options do not work</li> <li>• Observation</li> </ul>	
<b>Radiation therapy and/or systemic therapy</b>	<ul style="list-style-type: none"> <li>• Radiation therapy is not usually recommended for desmoid tumors in the abdomen or retroperitoneum</li> <li>• For systemic therapy, <a href="#">see Guide 8, 9, and 10</a></li> <li>• Adjuvant radiation therapy</li> </ul>	

### Observation

Observation is a period of testing to watch for tumor growth so that treatment can be started if needed. Observation might be an option for tumors that are stable and are not causing any symptoms.

### Surgery

Large tumors that are causing problems will be treated based on the location of the tumor, if it is causing pain, or if it interferes with movement. Surgery might not be considered if there is risk of serious complications.

Options include:

- Surgery
- Radiation therapy with or without systemic therapy

You may receive more treatment after primary treatment. If the tumor is all gone, you may start observation. If disease remains, you might have radiation therapy, systemic therapy, or more surgery.

### Radiation therapy

In general, radiation therapy is only recommended for desmoid tumors in the limbs (arms and legs), outer torso, or head and neck.

Radiation therapy is not recommended for desmoid tumors located in the abdomen or retroperitoneum. The peritoneum is the lining that surrounds the abdominal organs. The retroperitoneum is the area behind and outside of the peritoneum. It includes organs such as major blood vessels, adrenal glands, kidneys, pancreas, and bladder.

### Systemic therapy

There are different types of systemic therapy used to treat desmoid tumors. It might include non-steroidal anti-inflammatory drugs (NSAIDs) such as sulindac or celecoxib. Celecoxib can cause heart issues or put those with heart issues at greater risk for side effects. NSAIDs help reduce fever, swelling, pain, and redness.

For a list of systemic therapies used to treat desmoid tumors, [see Guide 31](#).

### Follow-up care

After surgery and any adjuvant treatment, you will be monitored for symptoms. You should receive rehabilitation if needed. This may include occupational or physical therapy.

## Guide 31 Systemic therapies: Desmoid tumors

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Sulindac or other NSAIDs, including celecoxib

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Tamoxifen

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Tamoxifen with sulindac

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Toremifene

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Methotrexate and vinblastine

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Low-dose interferon

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Doxorubicin-based regimens

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Imatinib

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Sorafenib

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Methotrexate and vinorelbine

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Liposomal doxorubicin

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Report any new or worsening symptoms to your doctor. A change in symptoms may be a sign of tumor growth. Follow-up tests will watch to see if the desmoid tumor is growing or has returned. Tests include medical history, physical exam, and imaging. These tests should be done every 3 to 6 months for 2 to 3 years, then every 6 to 12 months. [See Guide 32.](#)

### Guide 32

#### Follow-up testing: Desmoid tumors

Evaluation for rehabilitation such as occupational therapy and physical therapy

Medical history and physical exam every 3 to 6 months for 2 to 3 years, then every 6 to 12 months

Imaging with CT or MRI every 3 to 6 months for 2 to 3 years, then every 6 to 12 months

## Unresectable

### Treatment options

There are different reasons why a desmoid tumor can't be removed with surgery (unresectable). Sometimes, surgery is too dangerous based on the location or your overall health.

There are 5 treatment options for a desmoid tumor that is unresectable:

- Ablation
- Radiation therapy
- Systemic therapy
- Surgery may be considered if other options do not work
- Observation

#### Ablation

Ablation uses extreme cold or extreme heat to destroy desmoid tumors.

#### Radiation therapy

Radiation therapy is a treatment option, as long as there isn't risk of serious complications.

#### Systemic therapy

There are different types of systemic therapy used to treat desmoid tumors. It might include NSAIDs such as sulindac or celecoxib. Celecoxib can cause heart issues or put those with heart issues at greater risk for side effects. NSAIDs help reduce fever, swelling, pain, and redness.

For a list of systemic therapies used to treat desmoid tumors, [see Guide 31.](#)

NCCN recommends considering a clinical trial for unresectable desmoid tumors that progress on systemic therapy.

### Surgery

Surgery might be considered if other options do not work. Perhaps some of the tumor can be removed.

Since one goal of treatment is to maintain function, amputation is almost never considered.

### Observation

Observation is a period of testing to watch for tumor growth so that treatment can be started if needed. Observation might be an option for tumors that are stable and are not causing any symptoms.

### Follow-up care

Follow-up care will start after treatment is complete. You should receive rehabilitation if needed. This may include occupational or physical therapy. Report any new or worsening 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 a medical history, physical exam, and imaging. These tests should be done every 3 to 6 months for 2 to 3 years. If results are normal during this time, these tests may be done every 6 to 12 months. [See Guide 32.](#)

## Review

- Desmoid tumors are also known as aggressive fibromatoses.
- A biopsy is needed to diagnose a desmoid tumor.
- Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating desmoid tumors.
- A tumor that can be removed with surgery is called resectable. Even though a tumor is resectable, surgery might not be the first treatment option.
- Observation, ablation, radiation therapy, systemic therapy, and surgery are all possible treatment options.
- Follow-up care will include rehabilitation, if needed. This may include occupational or physical therapy.
- You will have tests to see how the tumor is responding to treatment. Tests include a medical history, physical exam, and imaging.

# 8

## Rhabdomyosarcoma

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78 Overview

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80 Pleomorphic RMS

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80 Non-pleomorphic RMS

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83 Review



Rhabdomyosarcoma (RMS) is a type of soft tissue sarcoma that starts in the cells that normally develop into skeletal muscle. Skeletal muscles are those that we can control or move. However, RMS can be found in other parts of the body. Together, you and your doctor will choose a treatment plan that is best for you.

## Overview

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

Before we are born, cells called rhabdomyoblasts begin to form. These cells eventually become skeletal muscle. Rhabdomyoblasts can also develop into RMS.

### Skeletal muscles

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





Because RMS starts in the rhabdomyoblasts, it is more common in children.

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

RMS has 3 subtypes:

- Embryonal (including botryoid and spindle cell variants)
- Alveolar (including a solid variant)
- Pleomorphic histologies

Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating RMS.

### Treatment for RMS

Treatment for RMS is divided into:

- Pleomorphic
- Non-pleomorphic, which include embryonal and alveolar subtypes

PET or PET/CT scan may be useful for staging. For treatment and testing recommendations, [see Guide 33](#).

## Guide 33 Testing and treatment for RMS

<b>Pleomorphic RMS</b>	PET or PET/CT scan may be useful for initial staging
	Recommend treating like soft tissue sarcoma
<b>Non-pleomorphic RMS (includes alveolar and embryonal)</b>	PET or PET/CT scan may be useful for initial staging
	Referral to center that has expertise in treating RMS is strongly recommended
	Multidisciplinary evaluation involving pediatric, medical, surgical, and radiation oncologists is strongly encouraged
	Treatment planning should include risk levels and combination of treatments

## Pleomorphic RMS

In pleomorphic RMS, the tumor is made up of more than one type of cell. Pleomorphic RMS occurs mostly in adults and tends to grow quickly. These tumors are commonly found in the limbs, torso wall, urinary system (kidneys, bladder, urethra, and ureter), and reproductive (vagina, uterus, testes, etc.) system.

### Treatment

Pleomorphic RMS is treated like soft tissue sarcoma. Treatment may involve surgery, radiation therapy, and systemic therapy. For a list of systemic therapies, see [Guide 34 and 35](#).

## Non-pleomorphic RMS

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

### Treatment

For non-pleomorphic RMS, including alveolar and embryonal types, treatment should be planned by a team of specialists at a center experienced in treating non-pleomorphic RMS. A pediatric oncologist may be involved in your care since this type is mostly seen in children. Medical, radiation, and surgical oncologists may also weigh in on your treatment options. Treatment planning should include risk groups and a combination of surgery, systemic therapy, and radiation therapy. For a list of systemic therapies, see [Guide 36 and 37](#).

**Guide 34****Combination systemic therapies: Pleomorphic RMS**

Doxorubicin and dacarbazine (called AD)

Doxorubicin, ifosfamide, and mesna (called AIM)

Mesna, doxorubicin, ifosfamide, and dacarbazine (called MAID)

Ifosfamide, epirubicin, and mesna

Gemcitabine and docetaxel

Gemcitabine and vinorelbine (for palliative therapy)

Gemcitabine and dacarbazine

**Guide 35****Single-agent systemic therapies: Pleomorphic RMS**

Doxorubicin

Ifosfamide

Epirubicin

Gemcitabine

Dacarbazine

Liposomal doxorubicin

Temozolomide (for palliative therapy)

Vinorelbine (for palliative therapy)

Eribulin (for palliative therapy)

Trabectedin (for palliative therapy)

Pazopanib (for palliative therapy)

Regorafenib (for non-adipocytic sarcoma)

Larotrectinib (for *NTRK* gene fusion-positive sarcomas)

Entrectinib (for *NTRK* gene fusion-positive sarcomas)

**Guide 36****Combination systemic therapies: Non-pleomorphic RMS**

Vincristine, dactinomycin, and cyclophosphamide

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Vincristine, doxorubicin, and cyclophosphamide

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Vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide

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Vincristine, doxorubicin, and ifosfamide

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Cyclophosphamide and topotecan

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Ifosfamide and doxorubicin

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Ifosfamide and etoposide

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Irinotecan and vincristine

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Vincristine and dactinomycin

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Carboplatin and etoposide

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Vinorelbine and low-dose cyclophosphamide (for palliative therapy)

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Vincristine, irinotecan, and temozolomide

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**Guide 37****Single-agent systemic therapies: Non-pleomorphic RMS**

Doxorubicin

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Irinotecan

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Topotecan

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Vinorelbine (for palliative therapy)

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High-dose methotrexate (in some cases)

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Trabectedin (for palliative therapy)

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## Review

- Rhabdomyosarcoma (RMS) is a rare type of sarcoma in adults. It is more common in children.
- RMS has 3 subtypes: embryonal, alveolar, and pleomorphic. Embryonal and alveolar are grouped into non-pleomorphic.
- Test results and treatment options should be reviewed by a team of experts from different fields of medicine who have experience in treating RMS.
- Those with RMS often require more than one type of cancer treatment including surgery, radiation therapy, and/or systemic therapy.
- Pleomorphic RMS is treated like soft tissue sarcoma. Treatment may involve surgery, radiation therapy, and systemic therapy.
- For non-pleomorphic RMS, including alveolar and embryonal types, treatment should be planned by a team of specialists at a center experienced in treating non-pleomorphic RMS.

# 9

## Making treatment decisions

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85 It's your choice

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85 Questions to ask your doctors

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92 Websites



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

## It's your choice

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

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

Some things that may play a role in your decisions:

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

Think about what you want from treatment. Discuss openly the risks and benefits of specific treatments and procedures. Weigh options and share concerns with your doctor. If you take the time to build a relationship with your doctor, it

will help you feel supported when considering options and making treatment decisions.

### Second opinion

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

### Things you can do to prepare

Check with your insurance company about its rules on second opinions. There may be out-of-pocket costs to see doctors who are not part of your insurance plan.

Make plans to have copies of all your records sent to the doctor you will see for your second opinion.

### Support groups

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

## Questions to ask your doctors

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

## Questions to ask about testing and staging

1. What tests will I have?
2. When will I have a biopsy? Will I have more than one? What are the risks?
3. Will I have any genetic tests?
4. How soon will I know the results and who will explain them to me?
5. Who will talk with me about the next steps? When?
6. What will you do to make me comfortable during testing?
7. Would you give me a copy of the pathology report and other test results?
8. What type of soft tissue sarcoma do I have? Where is it located? Has it spread?
9. What is the stage? What does this stage mean in terms of survival?
10. Can this soft tissue sarcoma be cured? If not, how well can treatment stop the cancer from growing?

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## Questions to ask about options

1. What will happen if I do nothing?
2. How do my age, overall health, and other factors affect my options?
3. What if I am pregnant? What if I'm planning to get pregnant in the near future?
4. Which option is proven to work best for my type of soft tissue sarcoma?
5. Does any option offer a cure or long-term cancer control? Are my chances any better for one option than another? Less time-consuming? Less expensive?
6. What are the possible complications and side effects?
7. Is surgery an option? Why or why not?
8. How do you know if treatment is working? How will I know if treatment is working?
9. What are my options if my treatment stops working?
10. What can be done to prevent or relieve the side effects of treatment?
11. Are there any life-threatening side effects of this treatment? How will I be monitored?
12. Can I stop treatment at any time? What will happen if I stop treatment?

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## Questions to ask about treatment

1. What are my treatment choices? What are the benefits and risks?
2. Which treatment do you recommend and why?
3. How long do I have to decide?
4. Will I have to go to the hospital or elsewhere for treatment? How often? How long is each visit? Will I have to stay overnight in the hospital or make travel plans?
5. Do I have a choice of when to begin treatment? Can I choose the days and times of treatment? Should I bring someone with me?
6. How much will the treatment hurt? What will you do to make me comfortable?
7. How much will this treatment cost me? What does my insurance cover? Are there any programs to help me pay for treatment?
8. Will I miss work or school? Will I be able to drive?
9. What type of home care will I need? What kind of treatment will I need to do at home?
10. When will I be able to return to my normal activities?
11. Which treatment will give me the best quality of life? Which treatment will extend my life? By how long?
12. I would like a second opinion. Is there someone you can recommend?

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## Questions to ask about surgery

1. What kind of surgery will I have? Will I have more than one surgery?
2. What will be removed during surgery?
3. How long will it take me to recover from surgery?
4. How much pain will I be in? What will be done to manage my pain?
5. What other side effects can I expect from surgery?
6. What treatment will I have before, during, or after surgery?

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## Questions to ask about clinical trials

1. What clinical trials are available for my type of soft tissue sarcoma?
2. What are the treatments used in the clinical trial?
3. What does the treatment do?
4. Has the treatment been used before? Has it been used for other types of cancer?
5. What are the risks and benefits of this treatment?
6. What side effects should I expect? How will the side effects be controlled?
7. How long will I be on the clinical trial?
8. Will I be able to get other treatment if this doesn't work?
9. How will you know the treatment is working?
10. Will the clinical trial cost me anything? If so, how much?

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## Questions to ask about side effects

1. What are the side effects of treatment?
2. How long will these side effects last?
3. What side effects should I watch for?
4. When should I call the doctor about my side effects? Can I text?
5. What medicines can I take to prevent or relieve side effects?
6. What can I do to help with pain and other side effects?
7. Will you stop treatment or change treatment if I have side effects?
8. What can I do to prevent side effects? What will you do?

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## Websites

### American Cancer Society

[cancer.org/cancer/soft-tissue-sarcoma.html](https://cancer.org/cancer/soft-tissue-sarcoma.html)

### Friends of TJ Foundation

[friendsoftj.org](https://friendsoftj.org)

### GIST Support International

[gistsupport.org](https://gistsupport.org)

### Leiomyosarcoma Support & Direct Research Foundation

[lmsdr.org](https://lmsdr.org)

### National Cancer Institute

[cancer.gov/cancertopics/pdq/treatment/adult-soft-tissue-sarcoma/patient](https://cancer.gov/cancertopics/pdq/treatment/adult-soft-tissue-sarcoma/patient)

### National Leiomyosarcoma Foundation

[nlmsf.org](https://nlmsf.org)

### Northwest Sarcoma Foundation

[nwsarcoma.org](https://nwsarcoma.org)

### QuadW Foundation (What Would Willie Want)

[quadw.org](https://quadw.org)

### Sara's Cure

[sarascure.org](https://sarascure.org)

### SARC

[sarctrials.org](https://sarctrials.org)

### Sarcoma Alliance

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

### Sarcoma Foundation of America

[curesarcoma.org](https://curesarcoma.org)

### Summer's Way Foundation

[summersway.org](https://summersway.org)

### The Alan B. Slifka Foundation

[slifkafoundation.org](https://slifkafoundation.org)

### The EHE Foundation

[fightehe.org](https://fightehe.org)

### The Life Raft Group

[liferaftgroup.org](https://liferaftgroup.org)

### The Paula Takacs Foundation for Sarcoma Research

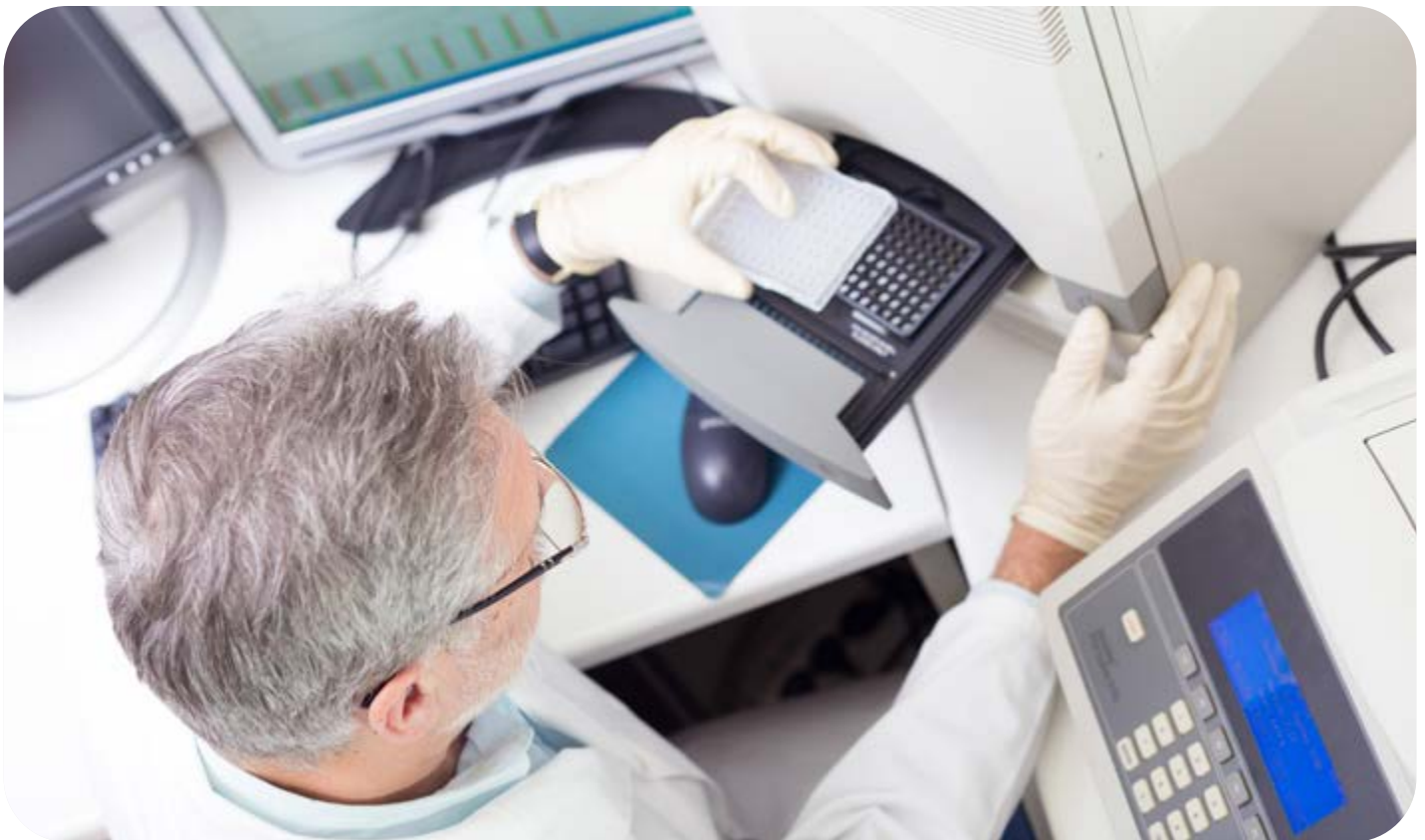
[paulatakacsfoundation.org](https://paulatakacsfoundation.org)

# 10

## Sarcoma mutations

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94 Sarcoma mutations



## Sarcoma mutations

Genetic tests might be useful for diagnosing and treating soft tissue sarcoma. Testing should be carried out by a pathologist who is experienced in sarcoma diagnosis and genetic testing techniques. The following guide is a list of some, but not all, of the mutations found in some types of soft tissue sarcoma.

### Guide 38 Malignant round cell tumor mutations

	Mutation	Gene(s) involved
Alveolar RMS	t(2;13)(q35;q14)	<i>PAX3-FOXO1</i>
	t(1;13)(p36;q14)	<i>PAX7-FOXO1</i>
	t(X;2)(q13;q35)	<i>PAX3-AFX</i>
Desmoplastic small round cell tumor	t(11;22)(p13;q12)	<i>EWSR1-WT1</i>
Embryonal RMS	Complex changes	Multiple, <i>MYOD1</i> mutation
Ewing sarcoma/peripheral neuroectodermal tumor	t(11;22)(q24;q12)	<i>EWSR1-FLI1</i>
	t(21;22)(q22;q12)	<i>EWSR1-ERG</i>
	t(2;22)(q33;q12)	<i>EWSR1-FEV</i>
	t(7;22)(p22;q12)	<i>EWSR1-ETV1</i>
	t(17;22)(q12;q12)	<i>EWSR1-E1AF</i>
	inv(22)(q12q;12)	<i>EWSR1-ZSG</i>
	t(16;21)(p11;q22)	<i>FUS-ERG</i>



Seek treatment at a center that has experience in your type of soft tissue sarcoma.

### Guide 39 Lipomatous tumor mutations

	Mutation	Gene(s) involved
<b>Atypical lipomatous tumor/well-differentiated liposarcoma (ALT/WDLS) and dedifferentiated liposarcoma</b>	Supernumerary ring chromosomes; giant marker chromosomes	Amplification of region 12q14-15, including <i>MDM2</i> , <i>CDK4</i> , <i>HMGA2</i> , <i>SAS</i> , <i>GL1</i>
<b>Myxoid/round cell liposarcoma</b>	t(12;16)(q13;p11)	<i>FUS-DD1T3</i>
	t(12;22)(q13;q12)	<i>EWSR1-DD1T3</i>
<b>Pleomorphic liposarcoma</b>	Complex changes	Unknown

**Guide 40**  
**Mutations in other sarcomas**

	<b>Mutation</b>	<b>Gene(s) involved</b>
<b>Alveolar soft part sarcoma</b>	der(17)t(X;17)(p11;q25)	<i>ASPL-TFE3</i>
<b>Angiomatoid fibrous histiocytoma</b>	t(12;22)(q13;q12)	<i>EWSR1-ATF1</i>
	t(2;22)(q33;q12)	<i>EWSR1-CREB1</i>
	t(12;16)(q13;p11)	<i>FUS-ATF1</i>
<b>Clear cell sarcoma</b>	t(12;22)(q13;q12)	<i>EWSR1-ATF1</i>
	t(2;22)(q33;q12)	<i>EWSR1-CREB1</i>
<b>Congenital/infantile fibrosarcoma</b>	t(12;15)(p13;q25)	<i>ETV6-NTRK3</i>
<b>Dermatofibrosarcoma protuberans</b>	t(17;22)(q21;q13) and derivative ring chromosomes	<i>COL1A1-PDGFB</i>
<b>Desmoid fibromatosis</b>	Trisomy 8 or 20; loss of 5q21	<i>CTNNB1</i> or <i>APC</i> mutations
<b>High-grade endometrial stromal tumors</b>	t(10;17)(q22;p13)	<i>YWHAE-FAM22A/B</i>
<b>Epithelioid hemangioendothelioma</b>	t(1;13)(p36;q25)	<i>WWTR1-CAMTA1</i>
	t(X;11)(q22;p11.23)	<i>YAP1 - TFE3</i>
<b>Epithelioid sarcoma</b>	Inactivation, deletion, or mutation of <i>INI1</i> ( <i>SMARCB-1</i> )	<i>INI1</i> ( <i>SMARCB-1</i> )
<b>Extrarenal rhabdoid tumor</b>	Inactivation of <i>INI1</i> ( <i>SMARCB-1</i> )	<i>INI1</i> ( <i>SMARCB-1</i> )

## Guide 41

## Mutations in other sarcomas (continued)

	Mutation	Gene(s) involved
Extraskeletal myxoid chondrosarcoma	t(9;22)(q22;q12)	<i>EWSR1-NR4A3</i>
	t(9;17)(q22;q11)	<i>TAF2N-NR4A3</i>
	t(9;15)(q22;q21)	<i>TCF12-NR4A3</i>
	t(3;9)(q11;q22)	<i>TFG-NR4A3</i>
Sporadic and familial GIST	Activating kinase mutations	<i>KIT</i> or <i>PDGFRA</i>
Carney-Stratakis syndrome (gastric GIST and paraganglioma)	Krebs cycle mutation	germline <i>SDH</i> subunit mutations
Inflammatory myofibroblastic tumor (IMT)	t(1;2)(q22;p23)	<i>TPM3-ALK</i>
	t(2;19)(p23;p13)	<i>TPM4-ALK</i>
	t(2;17)(p23;q23)	<i>CLTC-ALK</i>
	t(2;2)(p23;q13)	<i>RANBP2-ALK</i>
	t(2;11)(p23;p15)	<i>CARS-ALK</i>
	inv(2)(p23;q35)	<i>ATIC-ALK</i>
Leiomyosarcoma	Complex changes	Unknown
Low-grade fibromyxoid sarcoma	t(7;16)(q33;p11)	<i>FUS-CREB3L2</i>
	t(11;16)(p11;p11)	<i>FUS-CREB3L1</i>
Malignant peripheral nerve sheath tumor		<i>NF1</i> , <i>CDKN2A</i> and <i>EED</i> or <i>SUZ12</i>
Mesenchymal chondrosarcoma		<i>HEY1 - NCOA2</i>

## Guide 42

## Mutations in other sarcomas (continued)

	Mutation	Gene(s) involved
<b>Solitary fibrous tumor</b>		<i>NAB2 - STAT6</i>
<b>Synovial sarcoma</b>	t(X;18)(p11;q11)	<i>SS18-SSX1</i>
	t(X;18)(p11;q11)	<i>SS18-SSX2</i>
	t(X;18)(p11;q11)	<i>SS18-SSX4</i>
<b>Tenosynovial giant cell tumor/ pigmented villonodular synovitis (TGCT/PVNS)</b>	t(1;2)(p13;q35)	<i>CSF1</i>



## Words to know

**ablation**

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

**adjuvant therapy**

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

**angiogram**

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

**biopsy**

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

**brachytherapy**

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

**cancer stage**

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

**Carney-Stratakis syndrome**

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

**catheter**

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

**chemoembolization**

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

**chemoradiation**

Treatment with a combination of chemotherapy and radiation therapy.

**chemotherapy**

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

**computed tomography (CT)**

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

**contrast**

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

**core needle biopsy**

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

**cryotherapy**

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

**desmoid tumor**

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

**distant recurrence**

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

**embolization**

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

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

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

**external beam radiation therapy (EBRT)**

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

**familial adenomatous polyposis (FAP)**

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

**fascia**

A deep layer of soft tissue.

**fine-needle aspiration (FNA)**

Use of a thin needle to remove a small amount of tissue or fluid from the body to test for cancer cells.

**Gardner's syndrome**

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

**gastroenterologist**

A doctor who's an expert in digestive diseases.

**gene**

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

**gastrointestinal stromal tumor (GIST)**

Type of soft tissue sarcoma that usually begins in cells in the wall of the gastrointestinal tract.

**grade**

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

**hereditary**

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

**histology**

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

**immune system**

The body's natural defense against infection and disease.

**immunotherapy**

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

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

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

**intraoperative radiation therapy (IORT)**

Radiation therapy given during surgery.

**isolated limb infusion/perfusion**

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

***KIT* gene**

Provides instructions for cell growth.

**laparoscopic surgery**

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

**Li-Fraumeni syndrome**

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

**local recurrence**

Cancer that returns after treatment. Found in or near the same area.

**lymph node**

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

**lymph node dissection**

Surgery to remove one or more lymph nodes.

**magnetic resonance imaging (MRI)**

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

**medical oncologist**

A doctor who's an expert in cancer drugs.

**metastasectomy**

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

**metastasis**

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

**mutation**

An abnormal change.

**neoadjuvant treatment**

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

**observation**

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

**occupational therapist**

An expert in helping people live life unaided.

**oncology surgeon**

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

**palliative care**

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

**pathologist**

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

**physical therapist**

An expert in helping people move better.

**plastic surgeon**

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

**platelet-derived growth factor receptor alpha (PDGFRA) gene**

Provides instructions for cell growth.

**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 cancerous cells.

**progression**

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

**radiation oncologist**

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

**radiation therapy**

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

**radiofrequency ablation**

A treatment that destroys cancer cells by heating them with high-energy radio waves.

**radiologist**

A doctor who's an expert in imaging tests.

**recurrence**

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

**remission**

There are minor or no signs of disease.

**resectable**

Cancer that can be completely removed with surgery.

**retroperitoneum**

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

**rhabdomyosarcoma (RMS)**

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

**sarcoma**

A cancer of bone or soft tissue cells.



**stereotactic body radiation therapy (SBRT)**

Radiation therapy given in higher doses to smaller areas over 1 to 5 sessions of treatment.

**supportive care**

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

**surgical margin**

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

**targeted therapy**

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

**thoracic surgeon**

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

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

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

***TP53* gene**

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

**ultrasound**

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

**unresectable**

Cancer that can't be removed with surgery.

**wild-type GIST**

GIST that does not have *KIT* or *PDGFRA* mutations.

**x-ray**

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

# NCCN Contributors

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

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# NCCN Cancer Centers

Abramson Cancer Center  
at the University of Pennsylvania  
Philadelphia, Pennsylvania  
800.789.7366 • [pennmedicine.org/cancer](http://pennmedicine.org/cancer)

Fred & Pamela Buffett Cancer Center  
Omaha, Nebraska  
800.999.5465 • [nebraskamed.com/cancer](http://nebraskamed.com/cancer)

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

City of Hope National Medical 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  
[dfbwcc.org](http://dfbwcc.org)  
[massgeneral.org/cancer](http://massgeneral.org/cancer)

Duke Cancer Institute  
Durham, North Carolina  
888.275.3853 • [dukecancerinstitute.org](http://dukecancerinstitute.org)

Fox Chase Cancer Center  
Philadelphia, Pennsylvania  
888.369.2427 • [foxchase.org](http://foxchase.org)

Huntsman Cancer Institute  
at the University of Utah  
Salt Lake City, Utah  
877.585.0303  
[huntsmancancer.org](http://huntsmancancer.org)

Fred Hutchinson Cancer  
Research Center/Seattle  
Cancer Care Alliance  
Seattle, Washington  
206.288.7222 • [seattlecca.org](http://seattlecca.org)  
206.667.5000 • [fredhutch.org](http://fredhutch.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)

O'Neal Comprehensive  
Cancer Center at UAB  
Birmingham, Alabama  
800.822.0933 • [uab.edu/onealcancercenter](http://uab.edu/onealcancercenter)

Roswell Park Comprehensive  
Cancer Center  
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)

St. Jude Children's Research Hospital  
The University of Tennessee  
Health Science Center  
Memphis, Tennessee  
888.226.4343 • [stjude.org](http://stjude.org)  
901.683.0055 • [westclinic.com](http://westclinic.com)

Stanford Cancer Institute  
Stanford, California  
877.668.7535 • [cancer.stanford.edu](http://cancer.stanford.edu)

UC San Diego Moores Cancer Center  
La Jolla, California  
858.657.7000 • [cancer.ucsd.edu](http://cancer.ucsd.edu)

UCLA Jonsson  
Comprehensive Cancer Center  
Los Angeles, California  
888.662.8252 • 310.825.5268  
[cancer.ucla.edu](http://cancer.ucla.edu)

UCSF Helen Diller Family  
Comprehensive Cancer Center  
San Francisco, California  
800.689.8273 • [cancer.ucsf.edu](http://cancer.ucsf.edu)

University of Colorado Cancer Center  
Aurora, Colorado  
720.848.0300 • [coloradocancercenter.org](http://coloradocancercenter.org)

University of Michigan  
Rogel Cancer Center  
Ann Arbor, Michigan  
800.865.1125 • [rogelcancercenter.org](http://rogelcancercenter.org)

The University of Texas  
MD Anderson Cancer Center  
Houston, Texas  
800.392.1611 • [mdanderson.org](http://mdanderson.org)

University of Wisconsin  
Carbone Cancer Center  
Madison, Wisconsin  
608.265.1700 • [uwhealth.org/cancer](http://uwhealth.org/cancer)

UT Southwestern Simmons  
Comprehensive Cancer Center  
Dallas, Texas  
214.648.3111 • [utswmed.org/cancer](http://utswmed.org/cancer)

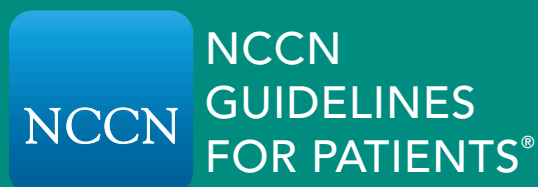
Vanderbilt-Ingram Cancer Center  
Nashville, Tennessee  
800.811.8480 • [vicc.org](http://vicc.org)

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

# Index

- ablation** 25
- adjuvant treatment** 49
- amputation** 23
- angiogram** 16
- brachytherapy** 25
- biopsy** 17, 22
- chemotherapy** 26
- chemoembolization** 25
- chemoradiation** 26
- clinical trial** 28
- computed tomography (CT)** 16
- embolization** 25
- external beam radiation therapy** 23-24
- familial adenomatous polyposis (FAP)** 18
- gene mutation** 61
- genetic testing** 18, 61
- grade** 19
- histology** 9
- immunotherapy** 26
- intraoperative radiation therapy (IORT)** 24
- isolated limb infusion/perfusion** 44
- KIT*** 62
- Li-Fraumeni syndrome** 18
- limb-sparing surgery** 23
- lymph node dissection** 48
- magnetic resonance imaging (MRI)** 16
- metastasectomy** 45
- neoadjuvant treatment** 49
- observation** 45
- PDGFRA*** 61
- palliative care** 56
- primary treatment** 51
- positron emission tomography (PET)** 16
- radiation therapy** 24-27
- radiofrequency ablation (RFA)** 25
- recurrence** 49
- rehabilitation** 23
- risk factors** 12
- staging** 19
- stereotactic radiation therapy (SBRT)** 24
- succinate dehydrogenase (SDH)** 18, 61
- supportive care** 28
- surgery** 22-23
- targeted therapy** 26-27
- TNM scores** 19
- ultrasound** 16
- wild-type GIST** 61
- x-ray** 16





# Soft Tissue Sarcoma

2020

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