

Soft Tissue Sarcoma Early Detection, Diagnosis, and Staging

Detection and Diagnosis

Catching cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that is not always the case.

- Can Soft Tissue Sarcomas Be Found Early?
- Signs and Symptoms of Soft Tissue Sarcomas
- How Are Soft Tissue Sarcomas Diagnosed?

Stages and Outlook (Prognosis)

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and anticipated response to treatment.

- Soft Tissue Sarcoma Stages
- Survival by Stage of Soft Tissue Sarcoma

Questions to Ask Your Cancer Care Team

Here are some questions you can ask your cancer care team to help you better understand your diagnosis and treatment options.

What Should You Ask Your Doctor About Soft Tissue Sarcomas?

Can Soft Tissue Sarcomas Be Found Early?

People who have a strong family history of sarcomas or who have had other cancers when they were young, might wish to discuss the benefits and disadvantages of genetic testing with their doctor. The test results should always be explained by a genetic counselor or a specially trained doctor who can interpret the results and advise high-risk patients about the need for early cancer detection tests.

Families with a history of certain inherited conditions (see What Are the Risk Factors for Soft Tissue Sarcomas?) caused by defects in certain genes have an increased risk of developing soft tissue sarcomas. The mutated genes can be detected by genetic testing, so family members should discuss this option with their doctors. They should let their doctor know about any lumps or growths right away.

No screening tests and exams are recommended for people who have no family history of sarcoma or other sarcoma risk factors. For these people, the best approach to early detection is to tell their health care professional about any unexplained lumps or growths or other symptoms that may be caused by a soft tissue sarcoma.

• References

See all references for Soft Tissue Sarcoma

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Signs and Symptoms of Soft Tissue Sarcomas

More than half of sarcomas begin in an arm or leg. Most people simply notice a **lump** that has grown over a period of time (weeks to months). Although the lump is often not painful, in some cases it will hurt.

When sarcomas grow in the back of the abdomen (the retroperitoneum), the symptoms they cause more often come from other problems. Sometimes the tumors cause pain. They may also cause blockage or bleeding of the stomach or bowels. They can grow large enough for the tumor to be felt in the abdomen. About 20% of sarcomas begin in the abdomen (stomach) area.

Sarcomas can also begin on the outside of the chest or abdomen (about 10%) or in the head or neck area (around 10%).

If you have any of the following problems, see a doctor right away:

- A new lump or a lump that is growing (anywhere on your body)
- Abdominal pain that is getting worse
- Blood in your stool or vomit
- Black, tarry stools (when bleeding happens in the stomach or bowels, the blood can turn black as it is digested, and it may make the stool very black and sticky)

These symptoms are more often caused by things other than sarcoma, but they still need to be checked out by a doctor.

References

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How Are Soft Tissue Sarcomas Diagnosed?

If you have signs or symptoms or other reasons to suggest you might have a sarcoma, your doctor will talk with you, examine you, and probably need to order some tests to find out if you have cancer.

Imaging tests

Some tests, such as a computed tomography (CT) scan or a magnetic resonance imaging (MRI) scan, are often done to look for the cause of symptoms and to find a tumor (such as a sarcoma). Other tests may be done after a sarcoma is diagnosed to look for cancer spread.

Plain x-ray

A regular x-ray of the area with the lump may be the first test ordered. A chest x-ray may be done after you are diagnosed to see if the sarcoma has spread to the lungs.

Computed tomography scans

The CT scan is an x-ray procedure that produces detailed, cross-sectional images of your body. Instead of taking one picture like a conventional x-ray, a CT scanner takes many pictures as it rotates around you. A computer then combines these pictures into an image of a slice of your body. The machine will create multiple images of the part of your body being studied. A CT scan is often done if the doctor suspects a soft tissue sarcoma in the chest, abdomen, or the retroperitoneum (the of the abdomen). This test is also used to see if the sarcoma has spread into the lungs, liver or other organs.

A CT scanner has been described as a large donut, with a narrow table in the middle opening. You will need to lie still on the table while the scan is being done. CT scans take longer than regular x-rays, and you might feel a bit confined by the ring while the pictures are being taken

Before any pictures are taken, you might be asked to drink 1 to 2 pints of a liquid called *oral contrast*. This helps outline the intestine more clearly. You may also receive an IV (intravenous) line through which a different kind of contrast dye (IV contrast) is injected. This helps better outline structures in your body.

The IV contrast dye can also cause some flushing (redness and warm feeling). Some people are allergic and get hives or, rarely, have more serious reactions like trouble breathing and low blood pressure. Be sure to tell the doctor if you have ever had a reaction to any contrast material used for x-rays.

CT scans might be done to precisely guide a biopsy needle into a tumor inside the body — the chest or abdomen, for example. For this procedure, called a *CT-guided needle biopsy*, the patient remains on the CT scanning table while a radiologist advances a biopsy needle toward the location of the mass. CT scans are repeated until the doctors are sure the needle is within the mass.

Magnetic resonance imaging scans

Magnetic resonance imaging (MRI) scans use radio waves and strong magnets instead of x-rays to take pictures of the body. The energy from the radio waves is absorbed and then released in a pattern formed by the type of tissue and by certain diseases. A

computer translates the pattern of radio waves given off by the tissues into a very detailed image of parts of the body. A contrast material might be injected, just as with CT scans, but is used less often.

MRI scans are often part of the work-up of any tumor that could be a sarcoma. They are often better than CT scans in evaluating sarcomas in the arms or legs. They provide a good picture of the extent of the tumor. They can show your health care team many things about the tumor, including location, size, and sometimes even the type of tissue it comes from (like fat or muscle). This makes MRI scans useful in planning a biopsy.

MRIs are also very helpful in examining the brain and spinal cord.

MRI scans are a little more uncomfortable than CT scans. First, they take longer — often up to an hour. Also, you have to lie inside a long tube, which is confining and can be upsetting. Special "open" MRI machines sometimes are an option for people who have claustrophobia (fear of enclosed spaces), but the drawback is that the pictures are often not as clear. MRI machines also make a thumping noise that you may find disturbing. Some places will provide headphones with music to block this noise out.

Ultrasound

Ultrasound uses sound waves and their echoes to produce pictures of parts of the body. A small instrument called a *transducer* emits sound waves and picks up the echoes as they bounce off the organs. A computer converts the sound wave echoes into an image that is displayed on a computer screen.

This is a very easy procedure to have. It uses no radiation, which is why it is often used to look at developing fetuses. For most ultrasounds, you simply lie on a table while a technician moves the transducer over the part of your body being examined. Usually, the skin is first lubricated with gel. Ultrasound may be done before a biopsy to see if a lump is a cyst, meaning if it has fluid and is likely benign, or if it is solid and more likely a tumor. This test is often not needed if a CT or MRI was done.

Positron emission tomography scan

In this test, radioactive glucose (sugar) is injected into the patient's vein to look for cancer cells. Because cancers use glucose (sugar) at a higher rate than normal tissues, the radioactivity will tend to concentrate in the cancer. A scanner can spot the radioactive deposits. A positron emission tomography (PET) scan is useful when your doctor thinks the cancer has spread but doesn't know where. A PET scan can be used instead of several different x-rays because it scans your whole body. Often the PET

scan is used with a CT scan. This helps decide if abnormalities seen on the CT scan are cancer or something else. PET is not often used for sarcoma, but it can be helpful in certain cases.

Biopsy

A biopsy is a procedure that removes a sample of tissue from a tumor to see if it is cancer. The piece of tissue is looked at under a microscope and, some other tests may be done on the sample as well. A physical exam or <u>imaging test</u> may suggest that a tumor is a sarcoma, but a biopsy is the only way to be certain that it is a sarcoma and not another type of cancer or a benign disease.

Several types of biopsies are used to diagnose sarcomas. Doctors experienced with these tumors will choose one, based on the size and location of the tumor. Most prefer to use a fine needle aspiration or a core needle biopsy as the first step.

Fine needle aspiration (FNA) biopsy

In FNA, the doctor uses a very thin needle and a syringe to withdraw small pieces of tissue from the tumor mass. The doctor can often aim the needle while feeling the mass near the surface of the body. If the tumor is too deep to feel, the doctor can guide the needle while viewing it on a computed tomography (CT) scan or ultrasound. The main advantage of FNA is that it can be used to biopsy tumors deep in the body without surgery. The disadvantage is that the thin needle may not remove enough tissue to make a precise diagnosis.

FNA is often useful in showing that a mass first thought to be a sarcoma (found on physical exam or imaging tests) is really another type of cancer, a benign tumor, an infection, or some other disease. But if FNA results suggest a sarcoma, another type of biopsy will usually be done to remove enough tissue to confirm that diagnosis. After a sarcoma is diagnosed, FNA is most useful in determining whether additional tumors in other organs are metastases.

Core needle biopsy

Core needle biopsies use a needle that is larger than the FNA needle. Sometimes this needle is called a *Tru-Cut* needle. It removes a cylindrical piece of tissue about 1/16 inch across and 1/2 inch long. It usually removes enough tissue to see if a sarcoma is present. Like FNA, CT scan and ultrasound can be used to guide the needle into tumors of internal organs.

Surgical biopsy

In a surgical biopsy, the entire tumor or a piece of the tumor is removed during an operation. There are 2 types of surgical biopsies, excisional and incisional. In an excisional biopsy, the surgeon removes the entire tumor. In an incisional biopsy, only a piece of a large tumor is removed. An incisional biopsy almost always removes enough tissue to diagnose the exact type and grade of sarcoma. If the tumor is near the skin surface, this is a simple operation that can be done with local or regional anesthesia (numbing medication given near the mass or into a nerve). But if the tumor is deep inside the body, general anesthesia is used (the patient is asleep).

If a tumor is rather small, near the surface of the body, and not located near critical tissues (such as important nerves or large blood vessels), the doctor may choose to remove the entire mass and a margin of normal tissue in an excisional biopsy. This surgery combines the biopsy and the treatment into one operation, so it should only be done by a surgeon with experience in treating sarcomas.

If the tumor is large, then an incisional biopsy is needed. Only a surgeon experienced in sarcoma treatment should perform this procedure.

You might want to ask about your surgeon's experience with this procedure. Proper biopsy technique is a very important part of successfully treating soft tissue sarcomas. An improper biopsy can lead to tumor spread and problems removing the tumor later on. An incisional biopsy in the wrong place or an excision without wide enough margins can make it harder to completely remove a sarcoma later on. To prevent these problems, these 2 types of biopsies should only be done by a surgeon experienced in treating sarcomas. It is best that an incisional biopsy be done by the same surgeon who will later remove the entire tumor (if a sarcoma is found).

Testing biopsy samples

The tissue removed will be looked at under the microscope to see if cancer is present. If it is, the doctor will try to determine what kind it is (sarcoma or carcinoma).

Grading: If a sarcoma is present, the biopsy will be used to determine what type it is and its grade. The grade of a sarcoma is based on how the cancer cells look under the microscope. In grading a cancer, the pathologist (a doctor who specializes in diagnosing diseases by looking at the tissue under a microscope) considers how closely the tumor resembles normal tissue (*differentiation*), how many of the cells appear to be dividing, and how much of the tumor is made up of dying tissue.

Each factor is given a score, and the scores are added to determine the grade of the tumor. Sarcomas that have cells that look more normal and have fewer cells dividing are generally placed in a low-grade category. Low-grade tumors tend to be slow growing, slower to spread, and often have a better outlook (prognosis) than higher-grade tumors. Certain types of sarcoma are automatically given higher differentiation scores. This affects the overall score so much that they are never considered low grade. Examples of these include synovial sarcomas and embryonal sarcomas.

The grade is partly used to determine the stage of a sarcoma. The official staging system (see How Are Soft Tissue Sarcomas Staged?) divides sarcomas into 3 grades (1 to 3). The grade of a sarcoma helps predict how rapidly it will grow and spread. It is useful in predicting a patient's outlook and helps determine treatment options.

Immunohistochemistry: Sometimes these special tests are needed to accurately determine whether a sarcoma is present and, if so, what type. Part of the biopsy sample is treated with special man-made antibodies that recognize cell proteins typical of certain kinds of sarcomas. The cells are treated with chemicals that make the cells containing these specific proteins change color. The color change is then seen under a microscope.

Cytogenetics: For this test, cells' chromosomes are examined with a microscope to look for changes. For example, in certain types of sarcomas part of one chromosome may be abnormally attached to part of a different chromosome (called a *translocation*). To see the chromosomes clearly, the cancer cells must be grown in laboratory dishes until they start dividing. This can take a week or more.

Fluorescent in situ hybridization (FISH) can sometimes be used to detect translocations and other chromosome changes without first growing the cells in the lab. Tests of chromosome changes are not required to diagnose a sarcoma, but they are sometimes very useful in confirming that a certain type of sarcoma is present. And as new changes are discovered, these tests may become more important and more common.

Reverse transcription polymerase chain reaction (RT-PCR): This test is another way to find translocations in some sarcomas (such as the Ewing family of tumors, alveolar rhabdomyosarcoma, and synovial sarcoma) to confirm the type of tumor. Instead of using a microscope to look for the chromosome changes as in cytogenetic testing or FISH, RT-PCR uses chemical analysis of the RNA (a substance that is made from DNA) from genes affected by the translocation. RT-PCR testing is often able to find translocations that aren't detected by cytogenetics.

References

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Soft Tissue Sarcoma Stages

After someone is diagnosed with a soft tissue sarcoma, doctors will try to figure out if it has spread, and if so, how far. This process is called **staging**. The stage of a cancer describes how much cancer is in the body. It helps determine how serious the cancer is and how best to <u>treat</u> it. Doctors also use a cancer's stage when talking about survival statistics.

The stages of soft tissue sarcomas range from stages I (1) through IV (4). As a rule, the lower the number, the less the cancer has spread. A higher number, such as stage IV, means cancer has spread more. And within a stage, an earlier letter means a lower stage. Although each person's cancer experience is unique, cancers with similar stages tend to have a similar outlook and are often treated in much the same way.

How is the stage determined?

The staging system most often used for soft tissue sarcomas is the American Joint Committee on Cancer (AJCC) **TNM** system, which is based on 4 key pieces of information:

- The extent of the tumor (T): How large is the cancer?
- The spread to nearby lymph nodes (N): Has the cancer spread to nearby lymph nodes?
- The spread (metastasis) to distant sites (M): Has the cancer spread to distant organs such as the lungs?
- The **g**rade (**G**) of the cancer: How much do the sarcoma cells look like normal cells?

The grade of a sarcoma is determined using a system known as the **French** or **FNCLCC system**, and is based on 3 factors:

- **Differentiation:** Cancer cells are given a score of 1 to 3, with 1 being assigned when they look similar to normal cells and 3 being used when the cancer cells look very abnormal. Certain types of sarcoma are given a higher score automatically.
- **Mitotic count:** How many cancer cells are seen dividing under the microscope; given a score from 1 to 3 (a lower score means fewer cells were seen dividing)
- **Tumor necrosis:** How much of the tumor is made up of dying tissue; given a score from 0 to 2 (a lower score means there was less dying tissue present).

The scores for each factor are added to determine the grade for the cancer. Higher-grade cancers tend to grow and spread faster than lower-grade cancers.

GX: The grade cannot be assessed (because of incomplete information).

Grade 1 (G1): Total score of 2 or 3

Grade 2 (G2): Total score of 4 or 5

Grade 3 (G3): Total score of 6, 7 or 8.

There are different staging systems for soft tissue sarcomas depending on where the cancer is in the body.

- Head and neck
- Trunk and extremities (arms and legs)
- Abdomen and thoracic (chest) visceral organs
- Retroperitoneum

Numbers or letters after T, N, and M provide more details about each of these factors. Higher numbers mean the cancer is more advanced. Once a person's T, N, and M categories have been determined, this information is combined in a process called **stage grouping** to assign an overall stage. Of the 4 main locations, only 2 (Trunk and Extremities and Retroperitoneum) have stage groupings. For more information see Cancer Staging.

The staging system in the table below uses the **pathologic stage** (also called the**surgical stage**). It is determined by examining tissue removed during an operation. Sometimes, if surgery is not possible right away or at all, the cancer will be given a **clinical** stage instead. This is based on the results of a physical exam, biopsy, and imaging tests. The clinical stage will be used to help plan treatment. Sometimes, though, the cancer has spread further than the clinical stage estimates, and may not predict the patient's outlook as accurately as a pathologic stage.

The system described below is the most recent AJCC system, effective January 2018. Cancer staging can be complex, so ask your doctor to explain it to you in a way you understand.

Trunk and Extremities Sarcoma Stages

AJCC	Stage	Trunk and Extremities Sarcoma
stage	grouping	Stage description* The cancer is 5 cm (2 inches) or
IA	T1 N0 M0 G1 or GX	The cancer is 5 cm (2 inches) or smaller (T1). It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 1 (G1) or the grade cannot be assessed (GX).
ΙΒ	T2, T3, T4 N0 M0 G1 or GX	The cancer is: • Larger than 5 cm but not more than 10 cm (T2) OR • Larger than 10cm but not more than 15 cm (T3) OR • Larger than 15 cm (T4). It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 1 (G1) or the grade cannot be assessed (GX). The cancer is 5 cm (2 inches) or
II	T1 N0 M0 G2 or G3	smaller (T1). It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 2 (G2) or grade 3 (G3).
IIIA	T2 N0 M0 G2 or G3	The cancer is larger than 5 cm (2 inches) but not more than 10 cm (T2). It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 2 (G2) or grade 3 (G3).
IIIB	T3 or T4 N0 M0 G2 or G3	The cancer is: • Larger than 10 cm but not more than 15 cm (T3) OR • Larger than 15 cm (T4).

		It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 2 (G2) or grade 3 (G3).
	M0 Any G	The cancer is any size (Any T) AND it has spread to nearby lymph nodes (N1). It has not spread to distant sites (M0). It can be any grade.
IV	OR	
	M1 Any G	The cancer is any size (Any T) AND it has spread to nearby lymph nodes (N1). It has spread to distant sites such as the lungs (M1). It can be any grade.

^{*}The following additional categories are not listed in the table above:

- TX: Main tumor cannot be assessed due to lack of information.
- **T0:** No evidence of a primary tumor.
- NX: Regional lymph nodes cannot be assessed due to lack of information.

Retroperitoneum Sarcoma Stages

AJCC	Stage	Retroperitoneum Sarcoma Stage
stage	grouping	description*
IA	T1 N0 M0 G1 or GX	The cancer is 5 cm (2 inches) or smaller (T1). It has not spread to nearby lymph nodes (N0) or to distant sites (M0). The cancer is grade 1 (G1) or the grade cannot be assessed (GX).
ΙΒ	T2, T3, T4 N0 M0 G1 or GX	 The cancer is: Larger than 5 cm but not more than 10 cm OR Larger than 10 cm but not more than 15 cm (T3) OR Larger than 15 cm (T4).

	1	Ilt has not appead to poorby lymph
		It has not spread to nearby lymph
		nodes (N0) or to distant sites (M0).
		The cancer is grade 1 (G1) or the
		grade cannot be assessed (GX).
		The cancer is 5 cm (2 inches) or
	T1	smaller (T1).
	N0	It has not spread to nearby lymph
II	мо	nodes (N0) or to distant sites (M0).
	G2 or G3	The cancer is grade 2 (G2) or grade 3
		(G3).
		The cancer is larger than 5 cm (2
	T2	inches) but not more than 10 cm (T2).
IIIA	NO	It has not spread to nearby lymph
	MO	nodes (N0) or to distant sites (M0).
	G2 or G3	` '
	GZ OF G3	The cancer is grade 2 (G2) or grade 3
		(G3). The cancer is:
		 Larger than 10 cm but not more
	T3 or T4	than 15 cm (T3) OR
	N0	 Larger than 15 cm (T4).
	MO	It has not spread to nearby lymph
	G2 or G3	nodes (N0) or to distant sites (M0).
шь		The cancer is grade 2 (G2) or grade 3
IIIB		(G3).
	OR	<u> </u>
		The cancer is any size (Any T) AND it
	Any T	has spread to nearby lymph nodes
	N1	(N1).
	MO	It has not spread to distant sites (M0).
	Any G	
		It can be any grade. The cancer is any size (Any T) AND it
IV	Any T	, , ,
	Any N	has spread to nearby lymph nodes
	M1	(N1).
	Any G	It has spread to distant sites such as
	[, •	the lungs (M1). It can be any grade.

^{*}The following additional categories are not listed in the table above:

- TX: Main tumor cannot be assessed due to lack of information.
- T0: No evidence of a primary tumor.
- NX: Regional lymph nodes cannot be assessed due to lack of information.

References

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See all references for Soft Tissue Sarcoma

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Survival by Stage of Soft Tissue Sarcoma

Survival rates are often used by doctors as a standard way of discussing a person's prognosis (outlook).

The 5-year survival rate (or *observed* survival rate) refers to the percentage of patients who live at least 5 years after their cancer is diagnosed. Of course, many people live much longer than 5 years (and many are cured).

Five-year *relative* survival rates assume that some people will die of other causes and compare the observed survival with that expected for people without the cancer. This is a better way to see the effect of the cancer on survival.

To get 5-year survival rates, doctors have to look at people who were treated at least 5 years ago. If treatment has improved since then, people now being diagnosed with soft tissue sarcoma may have a more favorable outlook.

Survival rates are often based on previous outcomes of large numbers of people who had the disease, but they cannot predict what will happen in any individual's case. Many other factors might affect a person's outlook, like the type of sarcoma, the location of the tumor, the treatment received, and the age of the patient. For example, sarcomas of the

arms or legs have a better outcome than those found in other places. Also, older patients tend to have worse outcomes than younger people. Your doctor can tell you how the numbers below may apply to you, as he or she is familiar with your particular situation.

The rates below are based on the stage of the cancer at the time of diagnosis. When looking at survival rates, it's important to understand that the stage of a cancer does not change over time, even if the cancer progresses. A cancer that comes back or spreads is still referred to by the stage it was given when it was first found and diagnosed, but more information is added to explain the current extent of the cancer. (And the treatment plan is adjusted based on the change in cancer status.)

The overall relative 5-year survival rate of people with soft tissue sarcomas is around 50% according to statistics from the National Cancer Institute (NCI). These statistics include people with Kaposi sarcoma, which has a poorer outlook than many sarcomas. The NCI doesn't use the AJCC staging system. Instead, they group sarcomas only by whether they are still confined to the primary site (called *localized*) have spread to nearby lymph nodes or tissues (called *regional*); or have spread (metastasized) to sites away from the main tumor (called *distant*). The 5-year survival rates for soft tissue sarcomas have not changed much for many years. The corresponding 5-year relative survival rates were:

- 83% for localized sarcomas (56% of soft tissue sarcomas were localized when they were diagnosed)
- 54% for regional stage sarcomas; (19% were in this stage)
- 16% for sarcomas with distant spread (16% were in this stage)

The 10-year relative survival rate is only slightly worse for these stages, meaning that most people who survive 5 years are probably cured.

For sarcomas of the arms and legs, Memorial Sloan-Kettering Cancer Center has survival rates broken down by AJCC stage (these are for observed, not relative survival):

Stone	5-year observed
Stage	survival rate
	90%
II	81%
Ш	56%
IV	Not available

Survival is worse when the sarcoma has developed somewhere other than the arms or legs. For example, the 5-year survival for retroperitoneal sarcomas is around 40% to

References

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What Should You Ask Your Doctor About Soft Tissue Sarcomas?

As you cope with cancer and cancer treatment, you need to have honest, open discussions with your doctor. You should feel comfortable asking any question no matter how small it might seem. Nurses, social workers, and other members of the treatment team may also be able to answer many of your questions.

- What kind of sarcoma do I have?
- How much experience do you have in diagnosing and treating sarcoma?
- Has my cancer spread?
- What is the <u>stage</u> of my cancer and what does that mean?
- What are my treatment choices?
- What treatment do you recommend and why?
- What risks or side effects are there to the treatments you suggest?
- What are the chances my cancer will come back with these treatment plans?
- What should I do to be ready for treatment?
- What's my outlook?

In addition to these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery times so that you can plan your work schedule. Or you may want to ask about second opinions or about <u>clinical</u> trials.

References

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