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Sarcoma Treatment Update

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Learn about:

- Different types of sarcomas
- Diagnosing sarcomas
- Treatment options
- Pain and symptom management



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For the best care, consult sarcoma center experts experienced in diagnosing and treating this rare type of cancer.

Sarcomas are a group of cancers of the bone or connective tissues such as fat, muscle, blood vessels, nerves, deep skin tissues, and cartilage. Sarcomas are rare, affecting less than one percent of adults with cancer and about 15 percent of children with cancer. But there are dozens of different types of sarcomas, and they can occur anywhere in the body.



One of the biggest challenges that doctors and patients face is trying to figure out exactly which bone and soft-tissue tumors are sarcomas and which are **benign** growths. That is why it is so important for people with sarcoma to seek advice at sarcoma treatment centers with experts who have extensive experience in diagnosing and treating this type of cancer. The Sarcoma Alliance posts a list of some of these centers on their website,

www.sarcomaalliance.org. You can also ask your doctor to refer you to a sarcoma treatment center.

Types of Sarcomas

There are hundreds of different types of sarcomas. Some of the most common types include:

- **Liposarcomas**, which develop in fat tissue.
- **Leiomyosarcomas**, which develop in smooth muscle tissue.
- **Rhabdomyosarcomas**, which form in a type of muscle called striated muscle.
- **Synovial sarcomas**, which develop in joints and appear more often in young adults.
- **Angiosarcomas**, which develop in the lining of the blood vessels.
- **Fibrosarcomas**, which develop in fibrous connective tissue found at the ends of bones in the arms or legs.
- **Malignant peripheral nerve sheath tumors**, which occur in the cells that surround nerves.
- **Gastrointestinal stromal tumors (GIST)**, which arise in the upper digestive tract.
- **Ewing's sarcomas**, which arise in soft tissue and bone.
- **Osteosarcomas**, which develop in bone.
- **Chondrosarcomas**, which develop in cartilage.

Diagnosing Sarcomas

Sarcomas often do not cause symptoms until the disease has become advanced. Most primary care doctors don't see many cases of sarcoma during their careers. That makes diagnosing these tumors particularly difficult.

Generally, anyone who has a persistent mass or lump that is getting bigger or is causing pain or other symptoms should consider a **biopsy**. A biopsy is a procedure in which some tissue from the tumor is removed in order to examine it under a microscope. A biopsy is the only way to determine whether a soft-tissue tumor is benign or **malignant** and, for sarcomas, what type of sarcoma it is. Three different types of

biopsy techniques are used to diagnose sarcomas:

Percutaneous biopsy The simplest and easiest biopsy technique is to put a small hollow needle through the skin into the tumor. The needle is used to withdraw cells or a small piece of tissue, which is examined under a microscope. Generally, percutaneous biopsy is the preferred technique because it is simple and 95 percent accurate in diagnosing sarcomas.

Excisional biopsy For this procedure, a surgeon cuts through the skin to remove the entire mass that is suspected of being cancerous.

Incisional biopsy This technique is similar to excisional biopsy, except that only a small part of the tumor is removed for examination. Both incisional and excisional biopsy techniques are surgical procedures that are done while



a patient is under local or general anesthesia. Generally, these types of biopsies are only performed in situations in which a percutaneous biopsy is not possible.

Once a sample has been taken, it has to be examined by a **pathologist**. Because sarcomas are so rare, review of the biopsy material by a pathologist who

has experience diagnosing sarcomas is very important. Whenever possible, this examination should be done before the patient receives any type of treatment.

Treatments for Sarcomas

Most localized sarcomas that have not spread to other parts of the body are treated with surgery to remove the tumor.

Your Medical Team

If you are diagnosed with sarcoma, one of the most important things you can do is work with a medical team that specializes in treating the disease. Ask your doctor to refer you to a sarcoma treatment center, whose health care professionals see hundreds of patients with sarcoma each year and who use a team approach to treating this cancer. If you prefer to keep working with your current oncologist, he or she can consult or work with a specialist team to make sure you get the best care possible.

To make communication with the team easier, it's helpful for you and your loved ones to choose a spokesperson who can communicate with all the members of the team and act as your advocate.

The sarcoma medical team will usually include the following health care professionals:

- **Pathologist** to examine biopsy material and other tissue samples
- **Diagnostic radiologist** to view the tumor
- **Surgical oncologist** to perform surgical procedures and limb-sparing surgery
- **Medical oncologist** to coordinate all aspects of cancer care
- **Radiation oncologist** to treat the cancer with radiation
- **Plastic surgeon** to improve appearance or reconstruct an arm or leg, if necessary
- **Orthopedic oncologist** to help with arm or leg reconstruction
- **Rehabilitation specialist** to help you regain any lost skills needed for daily living
- **Oncology nurses** to assist with all aspects of care
- **Psychiatrists, psychologists, and oncology social workers** to help you and your loved ones cope with the emotional and practical concerns of having cancer



Radiation is often added to surgery, and this combination has proven more effective than surgery alone, especially for deep tumors that are larger than five centimeters (about two inches) or those that cannot be removed with clean margins. A margin is the edge or border of the tumor tissue removed during cancer surgery. The margin is described as “clean” when there are no cancer cells at the edge of the tissue, suggesting that all of the cancer has been removed. Other treatments include chemotherapy, which is used to treat only certain types of sarcomas, and newer **targeted treatments**.

SURGERY

Significant advances in the surgical treatment of sarcomas have been made during the past three decades. In the past, people with sarcomas in the arms or legs usually had to undergo amputation—surgical removal—of those limbs as part of treatment. Now, these sarcomas can be treated with **limb-sparing surgery**. They are surgically removed in a way that preserves the use of the arm or the leg. The development of limb-sparing surgery has led to a dramatic drop in amputation rates among people with sarcoma, from about 50 percent in the past to less than five percent today.

Another surgical advance that has made a tremendous difference for people with sarcoma is the use of microvascular surgical techniques. When a sarcoma is removed, surgeons sometimes have to take a substantial amount of skin and muscle around the tumor as well, to make sure that all the sarcoma cells are taken away. But removing this tissue may cause changes to the appearance or full use of the arm, leg, or other area where the tumor had been. Microvascular surgical techniques allow surgeons to transfer muscle and skin from other parts of the body to the surgical site. This procedure helps the wound to heal properly and the surrounding area to maintain its function.

RADIATION

For many types of sarcomas, combining surgery with radiation treatment has proven more effective than treatment with surgery alone. There are several radiation techniques, as we describe below. In some cases, radiation is given after the tumor is surgically removed. Radiation treatments usually begin about four to six weeks after surgery and last for five to six-and-a-half weeks. Another option is to give radiation first and do surgery about four to eight weeks later.

Each approach—pre-operative radiation and post-operative radiation—has its advantages and disadvantages. For example, when radiation is given before surgery, doctors

can usually give a lower dose. In this way, the volume of tissue exposed to radiation is usually smaller. Giving radiation treatment before surgery also causes fewer long-term side effects than radiation given after surgery. Side effects from radiation can include fibrosis, a condition in



which the tissues can become very firm and inflexible, and edema, or swelling of the tissues, which can also become a long-term treatment side effect.

If a patient has surgery first, it's best to let the surgical wounds heal before radiation treatment is given. Radiation can impair the ability of some normal tissues to heal after surgery. Currently, no one approach is preferable. Each person should discuss the sequence of surgery and radiation treatment with his or her medical team.

Radiation treatments can be delivered in several different

ways. The most common is a type called external beam radiation, in which the radiation is delivered to the tumor from a machine outside the body. Another option, called brachytherapy, involves placing a radiation source inside the body, in direct contact with the tumor, for a short period after surgery. A third option is called intraoperative radiation. This type of radiation is delivered directly into the tumor at the time of surgery. It's important for patients to discuss their options thoroughly with their medical team. Which radiation technique doctors choose depends on the type of sarcoma, its location, and other factors.

Whether surgery alone or surgery combined with radiation is used, the goal of treatment is to remove the entire sarcoma with a margin of normal tissue around it while preserving as much function as possible. Occasionally, a sarcoma cannot be removed without either amputating a limb or causing a major decrease in its use. In such situations, radiation alone is sometimes used, to shrink the tumor and help control the cancer while preserving limb function.

CHEMOTHERAPY

Chemotherapy is used to treat three types of sarcomas, all of which tend to occur in children under the age of 18: osteosarcomas (bone sarcomas), Ewing's sarcomas (bone and soft-tissue sarcomas), and rhabdomyosarcomas (muscle sarcomas). Radiation may be combined with chemotherapy for effective treatment of these cancers. Chemotherapy alone is less effective at treating other types of sarcomas.

TARGETED TREATMENTS

Unlike chemotherapy, targeted treatments attack specific molecules and cell mechanisms thought to be important for cancer cell survival and growth. This specific targeting helps to spare healthy tissues and causes less severe side effects.

In 2002, the first successful targeted treatment, the drug

The Importance of Clinical Trials

There's no question that clinical trials have led to advances in cancer treatment, creating a brighter future for people with cancer. Clinical trials are the standard by which we measure the worth of new treatments and their impact on quality of life as patients go through those treatments. For this reason, doctors and scientists urge patients to take part.

Your doctor can guide you in making a decision about whether a clinical trial is right for you. Here are a few things you should know:

- Often, people who take part in clinical trials gain access to and benefit from new treatments.
- Before you participate in a clinical trial, you will be fully informed as to the risks and benefits of the trial.
- No patient receives a placebo (a look-alike pill or liquid with no active ingredient) if there is a standard treatment available for the disease. Most clinical trials are designed to test a new treatment against a standard treatment to find out whether the new treatment has any added benefit.
- You can stop taking part in a clinical trial at any time for any reason.

imatinib (Gleevec), was approved for use in people with a rare type of sarcoma in the upper digestive tract called gastrointestinal stromal tumor (GIST). In clinical trials, imatinib shrank tumors in about half of people with GIST. The tumors continued to grow in about 15 percent of patients who used imatinib. This medication works by preventing **receptors** from sending signals to cancer cells to grow and multiply. But after about two years of treatment with the drug, more than half the patients' tumors became **resistant** to imatinib—that is, the tumors continued growing despite treatment with the drug.

During the past several years, researchers have been studying different ways to treat imatinib-resistant GIST. The targeted treatment sunitinib (Sutent) was approved in 2006 for use in patients whose GIST has become resistant to imatinib. Sunitinib attacks several mechanisms in cancer cells, cutting off their blood supply and blocking their ability to grow.

Also, a newer promising targeted treatment, the oral form of a medication called ridaforolimus, is now being studied in SUCCEED, an international clinical trial. It is one of the largest studies of people with soft-tissue and bone sarcomas ever conducted and could change the way these patients are treated, researchers say. Ridaforolimus blocks a protein called mTOR, which is a master switch in cancer cells that signals them to grow and multiply. Patients and their loved ones should ask their doctors about this and other clinical trials on targeted treatments for sarcoma.

PAIN AND SYMPTOM MANAGEMENT

Sarcoma and the treatments for it can cause a number of side effects. A key to managing these side effects is to be aware of them and to communicate with your health care team when they arise. For example, it is important to discuss with your health care providers any pain you experience before, during, and after treatment. The type of pain and characteristics of pain may vary, and different strategies are used to manage different types of pain. It's helpful to score pain on a scale of zero to 10, where zero is no pain and 10 is the worst pain. By tracking the pain, you and your doctor can tell whether the recommended pain management techniques and treatments are working.

In addition, it's important to talk with your health care team about any quality-of-life concerns. As mentioned, sarcomas arise in many different areas of the body, and treatment strategies vary from person to person. For example, if a

sarcoma or its treatment will affect your ability to walk, it's important to discuss this with your health care team. Your doctor can then include rehabilitation specialists in your care plan.

Your Support Team

When you are diagnosed with sarcoma, you're faced with a series of choices that will have a major effect on your life, and maybe you're not sure where to turn. But help is available. Your health care team, family members, and friends will likely be an invaluable source of support at this time. You can also turn to these resources:

Oncology social workers provide emotional and practical support for people with cancer and their loved ones. These professionals can help you cope with the challenges of a sarcoma diagnosis and guide you to resources. CancerCare® offers free counseling from oncology social workers on staff who understand the challenges faced by people with sarcoma. We can work with you one-on-one to develop strategies for coping.

Support groups Many support groups are available for people with cancer. Support groups provide a caring environment in which you can share your concerns with others in similar circumstances.

Support group members come together to help one another, providing insights and suggestions on ways to cope. At CancerCare, people with sarcoma and their families can take part in support groups in person, online, or on the



telephone. All our groups are led by professional oncology social workers.

Financial help is offered by a number of organizations, including CancerCare®, to help cover cancer-related costs such as transportation to treatment, child care, or help needed around the home. CancerCare also provides referrals to other organizations that provide assistance.

To learn more about how CancerCare helps, call us at 1-800-813-HOPE (4673) or visit www.cancercares.org.

Frequently Asked Questions

Q My husband has a soft-tissue sarcoma. Are there any clinical trials that might be good for him?

A First, be sure that he gets an accurate diagnosis of the sarcoma and an expert opinion on treatment options from a sarcoma center. It may be that your husband has a good chance of doing well with standard treatments. If your oncologist believes that standard treatments will not be enough, he or she will likely give you information about clinical trials your husband may be eligible for. You can also get information about clinical trials from organizations such as the National Cancer Institute. Any information you find should be discussed with your husband's oncologist, who is familiar with the specifics of your husband's case and can let you know whether a particular clinical trial might be appropriate for him.

Q I just read a newspaper article about a new treatment for sarcoma. How can I tell whether it's a breakthrough treatment or something that's been hyped?

A This is a challenge for both doctors and patients, particularly with an unusual cancer like sarcoma. One thing that is helpful is to look for information from professional groups such as the National Cancer Institute's Cancer Information Service, the American Society of Clinical Oncology, or the other organizations listed on the Resources page at the end of this booklet. If the information is difficult to interpret, you can take it to your oncologist. He or she can help you understand it and determine whether a change in your treatment plan is appropriate based on the information.

Q A friend of mine suggested that I consider robotic surgery for my sarcoma. What exactly is this?

A Robotic surgery is a fairly new technique now being used in some hospitals. The technique allows a surgeon to perform an operation on a patient by using a device that is placed several feet away from the patient. The surgeon looks into a video monitor and uses a hand control to perform the surgery. The advantages of such surgery are thought to be better precision, smaller incisions, less blood loss, less pain, and quicker healing time. At this time, there are no open clinical trials of robotic surgery for sarcomas. Right now, surgery performed by a surgical oncologist at a sarcoma treatment center is still a better option than robotic surgery for patients with this type of cancer.

Glossary

benign Not cancerous; benign tumors may grow larger with time but do not spread to tissues around them or to other parts of the body.

biopsy Removal of a small piece of tissue, with either a hollow needle or surgery. The tissue is examined for cancer cells under a microscope.

limb-sparing surgery Surgically removing an arm or leg tumor in a way that preserves function of the limb.

malignant Cancerous; malignant tumors can spread to tissues surrounding them and may spread to other parts of the body.

pathologist A physician who examines cells and other tissues removed during biopsies to check for cancer and other diseases.

receptors Proteins located on the cancer cell's surface or inside the cell that serve as "doorways" for specific substances that encourage cancer cells to grow and divide.

resistant When cancer cells become resistant to a drug, they continue growing despite treatment with the drug.

sarcomas Cancers that arise in the bones and connective tissues, such as fat and muscle.

targeted treatments Unlike chemotherapy, targeted treatments attack specific molecules and cell mechanisms thought to be important for cancer cell survival and growth. This specific targeting helps to spare healthy tissues and causes less severe side effects.

Resources

CancerCare

Services: 1-800-813-HOPE (4673)
www.cancer.org

American Cancer Society

1-800-227-2345
www.cancer.org

Cancer.Net

Patient information from the American Society of Clinical Oncology
www.cancer.net

Cancer Support Community

www.cancersupportcommunity.org

National Cancer Institute

Cancer Information Service
1-800-422-6237
www.cancer.gov

National Coalition for Cancer Survivorship

See especially their Cancer Survival Toolbox®
1-877-650-9127
www.canceradvocacy.org

National Organization for Rare Disorders

1-203-744-0100
www.rarediseases.org

Sarcoma Alliance

1-415-381-7236
www.sarcomaalliance.org

Sarcoma Foundation of America

1-301-253-8687
www.curesarcoma.org

To find out about clinical trials:

Coalition of National Cancer Cooperative Groups

www.CancerTrialsHelp.org

National Cancer Institute

www.cancer.gov/clinicaltrials



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The information presented in this patient booklet is provided for your general information only. It is not intended as medical advice and should not be relied upon as a substitute for consultations with qualified health professionals who are aware of your specific situation. We encourage you to take information and questions back to your individual health care provider as a way of creating a dialogue and partnership about your cancer and your treatment.

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When one word changes your world,

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With CancerCare,
the difference comes from:

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