TREATMENT UPDATE:

Sarcoma

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Cancer*Care*®
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New York, NY 10001

Toll-free 800-813-HOPE (4673)
Fax 212-712-8495
Email info@cancercare.org
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Treatment Update: Sarcoma

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EDITOR

Breelyn A. Wilky, MD
Associate Professor, Director of Sarcoma Translational Research, Department of Medicine, Division of Medical Oncology, University of Colorado Anschutz Medical Campus

Sarcoma is a rare form of cancer, with approximately 15,000 cases diagnosed in the United States each year. There are two main types of sarcoma, each with many subtypes.

In soft tissue sarcoma, tumors begin in the tissues that connect and support other body structures, including muscle, fat, blood vessels, nerves, deep skin tissue and cartilage.

In osteosarcoma, tumors begin in the bones.

Soft tissue sarcoma accounts for approximately 90 percent of sarcoma cases.

Symptoms

The most common early sign of soft tissue sarcoma is a painless lump. The lump may get bigger and press against nerves or muscles, resulting in pain or discomfort. Weight loss and abdominal pain may occur in soft tissue sarcoma, although there are many other conditions that can cause these symptoms.

Early symptoms of osteosarcoma can include intermittent pain in the affected bone, which may be worse at night. The bone pain is sometimes accompanied by swelling and redness. A broken bone occurring unexpectedly (for example, as a result of a minor injury) may also be an early sign of osteosarcoma.

Diagnosis

Tests used to diagnose sarcoma include:

- **Physical examination.** A thorough physical exam will allow the doctor to better understand signs and symptoms, including any pain that is being experienced.
- **Imaging tests.** Ultrasound, PET scans, CT scans and/or MRIs are used to view connective tissue, and are helpful in diagnosing soft tissue sarcoma. Bone scans and X-rays are used if osteosarcoma is suspected.
- **Biopsy.** Using a hollow needle, the surgeon will remove a tissue sample from the tumor. The sample is then tested to determine if sarcoma cells are present.

Additional tests may be recommended once a diagnosis of sarcoma has been made, to determine if the cancer has spread to other parts of the body (metastasized).

Treatment Options

Surgery

Surgery is usually the primary approach in the treatment of sarcoma. The goal of surgery is to remove the tumor and as many cancer cells as possible without hurting nerves, organs or other important body structures.

Radiation

Intraoperative radiation therapy (IORT) may be administered during surgery to destroy sarcoma cells. IORT directs radiation to the tumor while sparing healthy surrounding tissue.

Radiation can also be administered before surgery to shrink the tumor (making the surgery easier) or after surgery to help prevent the cancer from recurring (coming back).

In cases where surgery is not possible, radiation therapy may be used to shrink the tumor. "External beam" radiation, in which a machine directs a beam (or multiple beams) of radiation to the tumor, is often used in treating sarcoma. Brachytherapy, in which radioactive material is placed inside the body, is another type of radiation therapy used to treat sarcoma.

Chemotherapy

Chemotherapy is the use of drugs to destroy cancer cells by stopping the ability of the cells to grow and divide. It is usually given into a vein using a needle (intravenously) or via a pill or capsule.

In the treatment of sarcoma, chemotherapy drugs can be used with or instead of surgery. Chemotherapy is often the first treatment choice if the sarcoma has metastasized.

The chemotherapy drugs most commonly used in the treatment of soft tissue sarcoma and osteosarcoma are ifosfamide (Ifex) and doxorubicin (Adriamycin). When ifosfamide is used, the drug mesna (Mesnex, others) is also given. Mesna reduces the risk of bleeding in the bladder, which is a potentially serious side effect of ifosfamide.

Other chemotherapy drugs used in the treatment of sarcoma include dacarbazine (DTIC), epirubicin (Ellence), temozolomide (Temoda), docetaxel (Taxotere), gemcitabine (Gemzar), vinorelbine (Navelbine), trabectedin (Yondelis) and eribulin (Halaven).

Chemotherapy drugs are typically used in combination for the treatment of sarcoma, as research has shown that this is more effective than monotherapy (a drug used alone).

Targeted therapy

Targeted therapies focus on specific molecules and cell mechanisms thought to be important for cancer cell survival and growth, taking advantage of what researchers have learned in recent years about how tumors grow.

In 2012, the U.S. Food and Drug Administration (FDA) approved pazopanib (Votrient) for the treatment of metastatic soft tissue sarcoma that was previously treated with chemotherapy. Pazopanib works by blocking the action of tyrosine kinases (proteins) that promote the formation of blood vessels, thereby "starving" the tumors.

Gastrointestinal stromal tumors (GISTs) are soft tissue sarcomas that can be located in any part of the digestive system. The FDA has approved two targeted therapies for the treatment of GISTs: imatinib (Gleevec) and sunitinib (Sutent). Both of these drugs interrupt the process of the spread of tumors.

Additional targeted therapy approaches are being studied in clinical trials.

Immunotherapy

Our immune system works constantly to keep us healthy. It recognizes and fights against danger, such as infections, viruses and growing cancer cells. In general terms, immunotherapy uses our own immune system as a treatment against cancer.

The immunotherapy denosumab (Xgeva, Prolia) is approved for the treatment of osteosarcoma. Denosumab blocks a factor in bone development known as RANK ligand, which stimulates cells that break bone down. By blocking RANK ligand, denosumab increases bone density and strength.

Immunotherapy as a treatment approach for sarcoma continues to be studied in clinical trials.

Ablation Therapy

Ablation therapy is a treatment that uses heat to destroy cancer cells. In radiofrequency ablation (RFA) and microwave ablation (MWA), the doctor uses an imaging technique, such as an ultrasound or CT scan, to guide a needle into the tumor. High-frequency electrical currents (in RFA) or microwaves (in MWA) are passed through the needle, creating a small region of heat that destroys the tumor.

The Importance of Clinical Trials

Clinical trials are the standard by which we measure the worth of new treatments and the quality of life of patients as they receive those treatments. For this reason, doctors and researchers urge people with cancer to take part in clinical trials.

Your doctor can guide you in making a decision about whether a clinical trial is right for you. Here are a few things that 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, including any possible side effects.
- 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.

Treatment Side Effects

All cancer treatments can cause side effects. It's important that you report any side effects that you experience to your health care team so they can help you manage them. Report them right away—don't wait for your next appointment. Doing so will improve your quality of life and allow you to stick with your treatment plan. It's important to remember that not all patients experience all side effects, and patients may experience side effects not listed here.

Side Effects of Radiation Therapy

Changes to the skin are the most common side effects of radiation therapy. Those changes can include dryness, swelling, peeling, redness and blistering. If a reaction occurs, contact a member of your health care team so the appropriate treatment can be prescribed. It's especially important to contact your health care team if there is any open skin or are any painful areas, as this could be sign of an infection. Infections can be treated with an oral antibiotic or topical antibiotic cream.

Side Effects of Chemotherapy

The side effects of chemotherapy depend on the type and dose of drugs given and the length of time they are used, and can include:

- Fatigue
- Nausea or vomiting
- Hair loss
- Increased risk of infection (from having too few white blood cells)
- Easy bruising or bleeding
- Changes in memory or thinking
- Peripheral neuropathy (numbness or tingling in hands and feet)

Side Effects of Targeted Therapy

Targeted therapy doesn't have the same effect on the body as do chemotherapy drugs, but they can still cause side effects. Side effects of targeted therapies can include diarrhea, liver problems (such as hepatitis and elevated liver enzymes), nerve damage, rashes, high blood pressure and problems with blood clotting and wound healing.

Side Effects of Immunotherapy

Immunotherapy travels through the bloodstream, helping to prompt an immune response. Sometimes the immune system may attack healthy cells as well as cancer cells, and certain side effects may be experienced, including fatigue, decreased appetite and digestive tract symptoms. The management of these potential side effects is discussed later in this booklet.

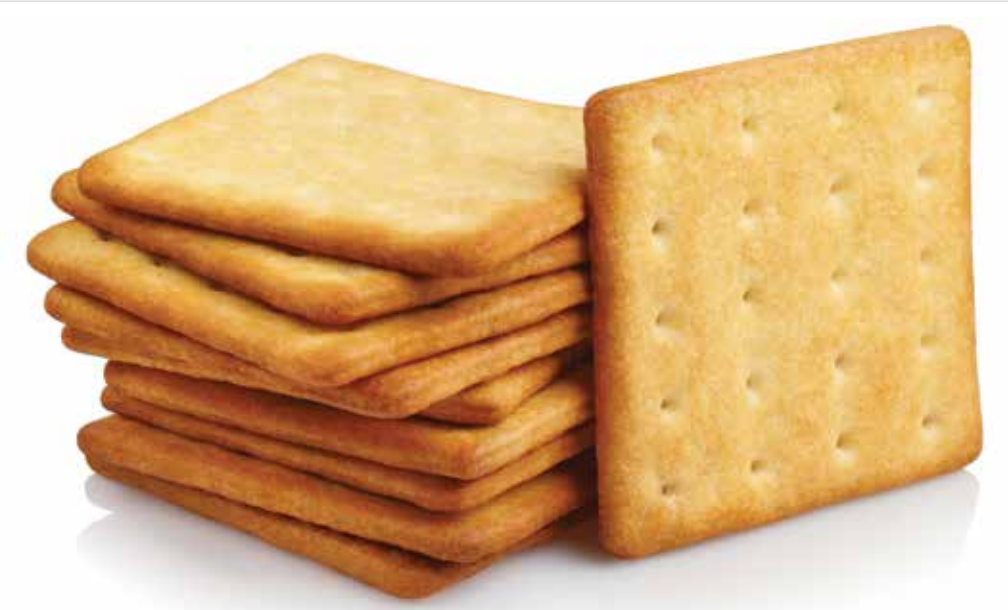
General Side Effects

Some side effects may occur across treatment approaches. This section provides tips and guidance on how to manage these side effects should they occur.

Managing Digestive Tract Symptoms

Nausea and vomiting

- Avoid food with strong odors as well as overly sweet, greasy, fried or highly seasoned food.
- Eat meals cold or at room temperature, which often makes food more easily tolerated.
- Nibble on dry crackers or toast. These bland foods are easy on the stomach.
- Having something in your stomach when you take medication may help ease nausea.



Diarrhea

- Drink plenty of water. Ask your doctor about using drinks such as Gatorade that provide electrolytes. Electrolytes are body salts that must stay in balance for cells to work properly.
- Over-the-counter medicines such as loperamide (Imodium A-D and others) and prescription drugs are available for diarrhea but should be used only if necessary. If the diarrhea is bad enough that you need medicine, contact a member of your health care team.
- Choose foods that contain soluble fiber, like beans, oat cereals and flaxseed, and high-pectin foods such as peaches, apples, oranges, bananas and apricots.
- Avoid foods high in refined sugar and those sweetened with sugar alcohols such as sorbitol and mannitol.

Loss of appetite

- Eating small meals throughout the day is an easy way to take in more protein and calories, which will help maintain your weight. Try to include protein in every meal.
- To keep from feeling full early, avoid liquids with meals or take only small sips (unless you need liquids to help swallow). Drink most of your liquids between meals.
- Keep high-calorie, high-protein snacks on hand such as hard-boiled eggs, peanut butter, cheese, ice cream, granola bars, liquid nutritional supplements, puddings, nuts, canned tuna or trail mix.
- If you are struggling to maintain your appetite, talk to your health care team about whether appetite-building medication could be right for you.

Managing Fatigue

Fatigue (extreme tiredness not helped by sleep) is one of the most common side effects of many cancer treatments. If you are taking a medication, your doctor may lower the dose of the drug, as long as it does not make the treatment less effective. If you are experiencing fatigue, talk to your doctor about whether taking a smaller dose is right for you.

There are a number of other tips for reducing fatigue:

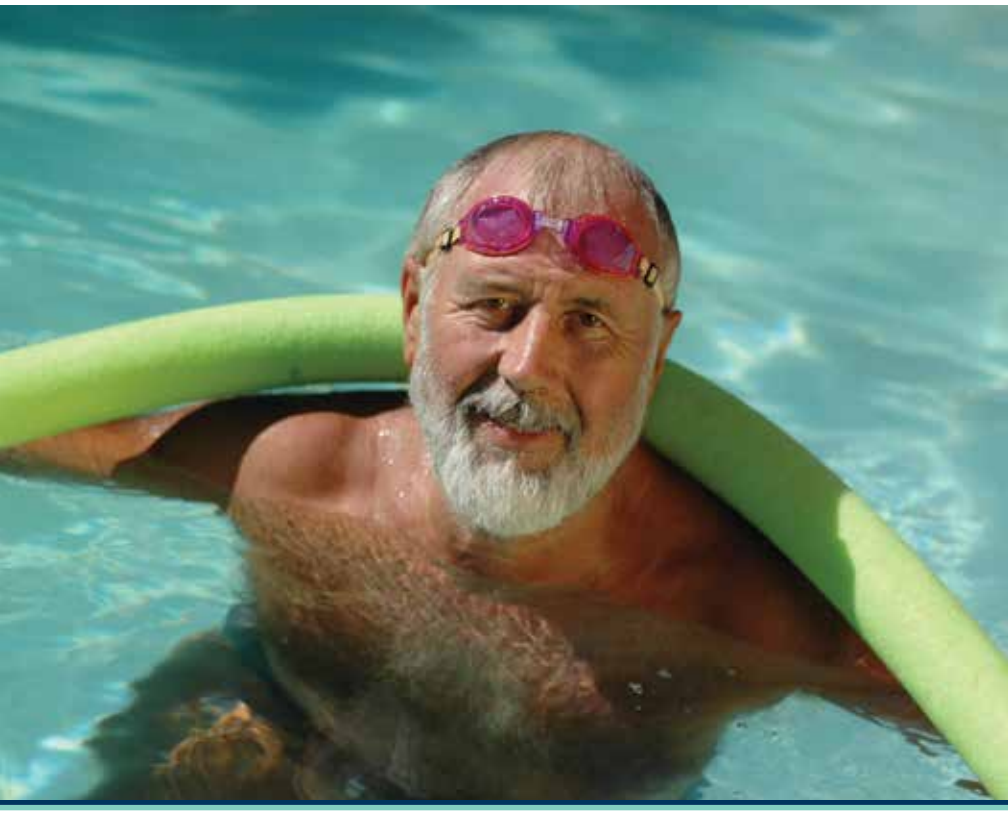
- Take several short naps or breaks during the day.
- Take walks or do some light exercise, if possible.
- Try easier or shorter versions of the activities you enjoy.
- Ask your family or friends to help you with tasks you find difficult or tiring.

There are also prescription medications that may help, such as modafinil. Your health care team can provide guidance on whether medication is the right approach for your individual circumstances.

Managing Pain

There are a number of options for pain relief, including prescription and over-the-counter medications. It's important to talk to a member of your health care team before taking any over-the-counter medication to determine if it is safe and to make sure it will not interfere with your treatment. Many pain medications can lead to constipation, which may make your pain worse. Your doctor can prescribe medications that help to avoid constipation.

Physical therapy, acupuncture and massage may also be of help in managing your pain. Consult with a member of your health care team before beginning any of these activities.



Communicating With Your Health Care Team

As you manage your sarcoma, it's important to remember that you are a consumer of health care. The best way to make decisions about health care is to educate yourself about your diagnosis and get to know the members of your health care team, including doctors, nurses, nurse practitioners, physician assistants, dietitians, social workers and patient navigators.

Here are some tips for improving communication with your health care team:

Start a health care journal. Having a health care journal or notebook will allow you to keep all of your health information in one place. You may want to write down the names and contact information of the members of your health care team, as well as any questions for your doctor.

Prepare a list of questions. Before your next medical appointment, write down your questions and concerns. Because your doctor may have limited time, ask your most important questions first and be as specific as possible.

Bring someone with you to your appointments. Even if you have a journal and a prepared list of questions or concerns, it's always helpful to have support when you go to your appointments. The person you bring may also think of questions to ask your doctor or remember details about your symptoms or treatment that you may have forgotten.

Write down your doctor's answers. Taking notes will help you remember your doctor's responses, advice and instructions. You can also ask the person who accompanies you to take notes for you, either in your journal or on a tablet or smartphone.

Record your visit if your doctor allows it. Recording the conversation with your doctor gives you a chance to hear specific information again or share it with family members or friends.

Incorporate other health care professionals into your team. Your medical oncologist is an essential member of your health care team, but there are other health care professionals who can help you manage your diagnosis and treatment:

- Your primary care physician should be kept updated about your cancer treatment and any test results.
- Your local pharmacist is a great source of knowledge about the medications you are taking. Have all of your prescriptions filled at the same pharmacy to avoid the possibility of harmful drug interactions.
- Make sure your oncologist knows of any other medical conditions you have or any pain you are experiencing so that they can consult with your primary care physician or specialist as needed.

Remember, there is no such thing as over-communication.

Cancer*Care*'s Free Support Services and Programs

It can be very difficult to receive a diagnosis of cancer, and adjusting to the necessary changes in your life can be challenging.

Cancer*Care* can help. We are a national nonprofit organization providing free, professional services to anyone affected by cancer. Our licensed oncology social workers can provide support and education, help in navigating the complicated health care system and offer information on support groups and other resources.

To learn more about how Cancer*Care* helps, call us at 800-813-HOPE (4673) or visit www.cancercare.org.

You will likely also build your own personal support network composed of family and friends. In doing so, it's best to take some time to think about the people in your life and how they are best suited to help. Match the task to their strengths—ask a family member who loves to shop to pick up something for you at the store, or ask a friend who's a good listener to come over for a chat.

Frequently Asked Questions

Q: Does it matter at what hospital or health care facility my treatment takes place?

A: Sarcoma is rare, and experts at sarcoma treatment centers have the latest information and extensive experience in diagnosing and treating this type of cancer. A list of some of these centers can be found on the Sarcoma Alliance website, www.sarcomaalliance.org. Your doctor may also be able to refer you to a nearby sarcoma treatment center.

Q: What is a treatment summary and why is important?

A: A treatment summary, sometimes called a "shadow chart," is a document that you create and keep in your possession. Maintaining your own records allows you and your family members to have instant access to the specifics of your sarcoma diagnosis and treatment. A treatment summary should include:

- Your name and date of birth
- Date of diagnosis
- Prescribed therapy/therapies; including dates started and stopped and dosages when appropriate
- Dates and types of post-diagnosis testing, and the results of these tests
- Other medications and supplements you are taking
- Names, affiliations and contact information of all members of your health care team

Talk to your doctor or a member of your health care team about your intention to create a treatment summary, and ask what else they suggest be included. Take your treatment summary with you when you visit any doctor, not just your oncologist.

Q: Are there factors that increase the risk of sarcoma?

A: While it is not known what causes sarcoma, there are some factors that increase the risk of it developing:

- **Inherited conditions.** Syndromes that increase the risk of soft tissue sarcoma include hereditary retinoblastoma, neurofibromatosis, Li-Fraumeni syndrome, tuberous sclerosis and Werner syndrome.
- **Radiation therapy.** Radiation treatment for cancer increases the risk of later developing sarcoma.
- **Lymphedema.** Lymphedema is swelling caused by a buildup of lymph fluid occurring when the lymphatic system is blocked or damaged. Lymphedema increases the risk of a type of sarcoma called angiosarcoma.
- **Exposure to viruses.** In people with a weakened immune system, the "human herpesvirus 8" virus can increase the risk of Kaposi's sarcoma, a type of sarcoma that forms in the lining of blood and lymph vessels.
- **Exposure to chemicals.** Long-term, high-dose exposure to certain chemicals such as herbicides, chlorophenols, arsenic, vinyl chloride and dioxin can increase the risk of a type of sarcoma that affects the liver.

Resources

CancerCare®
800-813-HOPE (800-813-4673)
www.cancercare.org

American Cancer Society
800-227-2345
www.cancer.org

Cancer.Net
Patient information from the American Society of Clinical Oncology
888-651-3038
www.cancer.net

National Cancer Institute
800-422-6237
www.cancer.gov

CLINICAL TRIALS WEBSITES

EmergingMed
www.emergingmed.com

National Cancer Institute
www.cancer.gov

Cancer Support Community
888-793-9355
www.cancersupportcommunity.org

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