

# **Treating Osteosarcoma**

If you or your child has been diagnosed with osteosarcoma, the cancer care team will discuss treatment options with you. It's important to weigh the benefits of each option against the possible risks and side effects.

#### How is osteosarcoma treated?

Three main types of treatment are used for osteosarcoma:

- Surgery for Osteosarcoma
- Chemotherapy and Other Drugs for Osteosarcoma
- Radiation Therapy for Osteosarcoma

#### **Common treatment approaches**

Most often, chemotherapy is given both before and after surgery. It can help lower the risk that the cancer will come back after urgery fo 0 rg /GS19F1 12 .7on

at a major cancer center. Doctors on the treatment team might include:

- An **orthopedic surgeon** (a surgeon who specializes in muscles and bones) who is experienced in treating bone tumors
- A **medical or pediatric oncologist** (a doctor who treats cancer with chemotherapy and other drugs)
- A radiation oncologist (a doctor who treats cancer with radiation therapy)
- A physiatrist (a doctor specializing in rehabilitation and physical therapy)

For both adults and children, the team might also include other doctors, physician assistants (PAs), nurse practitioners (NPs), nurses, psychologists, social workers, rehabilitation specialists, and other health professionals.

- Health Professionals Associated with Cancer Care
- How to Find the Best Cancer Treatment for Your Child
- Navigating the Health Care System When Your Child Has Cancer

#### Making treatment decisions

Treatment for osteosarcoma is often effective, but it can also cause serious side effects. It's important to discuss all treatment options as well as their possible side effects with the cancer care team so you can make an informed decision. It's also very important to ask questions if you're not sure about anything.

If time allows, getting a second opinion from another doctor experienced in treating osteosarcoma is often a good idea. This can give you more information and help you feel more confident about the treatment plan you choose. If you aren't sure where to go for a second opinion, ask your doctor for help.

Once treatment starts, members of the treatment team can help you deal with side effects, stress, and financial and other issues related to treatment.

For cancer in children and teens, many of these issues can be more complex. As a parent, taking care of a child with cancer can be a very big job. It's important to remember that you will have a lot of help. Many people will be involved in your child's overall care. It's also important for you to know that the health professionals who treat children with osteosarcoma are using the experience and knowledge gained from many decades of studying the treatment of this disease.

#### **Preparing for treatment**

Before treatment, the doctors and other members of the team will help you understand the tests that will need to be done. The team's social worker will also counsel you about some of the issues that might come up during and after treatment, and might be able to help you find housing and financial aid if needed.

• When Your Child Has Cancer

#### Help getting through cancer treatment

Your cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services can also be an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help. For children and teens with cancer and their families, other specialists can be an important part of care as well.

The American Cancer Society also has programs and services – including rides to treatment, lodging, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists.

- Finding Help and Support When Your Child Has Cancer
- Find Support Programs and Services in Your Area

The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask your cancer care team any questions you may have about your treatment options.

# Surgery for Osteosarcoma

Surgery is an important part of treatment for almost all osteosarcomas. It includes:

- The <u>biopsy<sup>1</sup></u> to diagnose the cancer
- The removal of the tumor(s)

Whenever possible, it's very important that the biopsy and the surgery to remove the tumor be planned together, and that an experienced orthopedic surgeon does both the biopsy and the surgery to remove the tumor. The biopsy should be done in a certain way to give the best chance that less extensive surgery will be needed later on.

The main goal of surgery is to remove all of the cancer. If even a small amount of cancer is left behind, it might continue to grow and make a new tumor, and might even spread to other parts of the body. To lower the risk of this happening, surgeons remove the tumor plus some of the normal tissue that surrounds it. This is known as a **wide excision**.

A doctor called a pathologist will look at the removed tissue under a microscope to see if there are cancer cells at the margins (outer edges).

- If cancer cells are seen at the edges of the tissue, the margins are called **positive**. Positive margins can mean that some cancer was left behind.
- When no cancer cells are seen at the edges of the tissue, the margins are said to be **negative**, **clean**, or **clear**. A wide excision with clean margins helps limit the risk that the cancer will come back in the place where it started.

The type of surgery done depends mainly on the location and size of the tumor. Although all operations to remove osteosarcomas are complex, tumors in the limbs (arms or legs) are generally not as hard to remove as those in the jaw bone, at the base of the skull, in the spine, or in the pelvic (hip) bone.

## Tumors in the arms or legs

Tumors in the arms or legs might be treated with either:

- Limb-salvage (limb-sparing) surgery: removing the cancer and some surrounding normal tissue but leaving the limb basically intact
- Amputation: removing the cancer and all or part of an arm or leg

Limb-salvage surgery

Most patients with tumors in the arms or legs can have limb-sparing surgery, but this depends on where the tumor is, how big it is, and whether it has grown into nearby structures.

Limb-salvage surgery is a very complex operation. The surgeons who do this type of operation must have special skills and experience. The challenge is to remove the entire tumor while still saving the nearby tendons, nerves, and blood vessels to keep as much of the limb's function and appearance as possible. If the cancer has grown into these structures, they will need to be removed along with the tumor. In such cases, amputation may sometimes be the best option.

The section of bone that is removed along with the osteosarcoma is replaced with a piece of bone from another part of the body or from another person (a **bone graft**) or with a man-made device made of metal and other materials that replaces part or all of a bone (an **internal prosthesis**). Some newer devices combine a graft and a prosthesis.

**Complications of limb-salvage surgery** can include infections and grafts or rods that become loose or broken. Patients who have limb-salvage surgery might need more surgery in the following years, and some might still eventually need an amputation.

Using an internal prosthesis in growing children is especially challenging. In the past, it required occasional operations to replace the prosthesis with a longer one as the child grew. Newer prostheses have become very sophisticated and often can be made longer without any extra surgery. They have tiny devices in them that can lengthen the prosthesis when needed to make room for a child's growth. But even these prostheses may need to be replaced with a stronger adult prosthesis once the child's body stops growing.

It takes about a year, on average, for patients to learn to walk after limb-salvage surgery on a leg. Physical rehabilitation after limb-salvage surgery is more intense than after amputation, and it's extremely important. If the patient doesn't actively take part in the rehabilitation program, the salvaged arm or leg might become useless.

#### Amputation

For some patients, amputation may be the best option. For example, if the tumor is very large or if it extends into the nerves and/or the blood vessels, it might not be possible to save the limb.

The surgeon determines how much of the arm or leg needs to be amputated based on the results of  $\underline{MRI \ scans}^2$  and an examination of removed tissue by the pathologist during the surgery.

Surgery is usually planned so that muscles and the skin will form a cuff around the remaining bone. This cuff will fit into the end of a prosthetic (artificial) limb. Another option might be to implant a prosthesis into the remaining bone, with the end of the

Emotional issues can be very important, and support and encouragement are needed for all patients. (See <u>Living as an Osteosarcoma Survivor</u><sup>3</sup>.)

# Tumors that start in other areas

**Tumors in the pelvic (hip) bones** can often be hard to remove completely with surgery. But if the tumor responds well to chemotherapy first, surgery (sometimes followed by radiation therapy) may get rid of all of the cancer. Pelvic bones can sometimes be reconstructed after surgery, but in some cases pelvic bones and the leg they are attached to might need to be removed.

For **tumors in the lower jaw bone**, the entire lower half of the jaw may be removed and later replaced with bones from other parts of the body. If the surgeon can't remove all of the tumor, radiation therapy may be used as well.

For **tumors in areas like the spine or the skull**, it may not be possible to remove all of the tumor safely. Cancers in these bones may require a combination of treatments such as chemotherapy, surgery, and radiation.

**Joint fusion (arthrodesis):** Sometimes, after the removal of a tumor that involves a joint (an area where two bones come together), it might not be possible to reconstruct the joint. In this case, surgery might be done to fuse the two bones together. This is most often used for tumors in the spine, but it might also be used in other parts of the body, such as a shoulder or hip. While it can help stabilize the joint, the person will have to learn to compensate for the resulting loss of motion.

## Surgical treatment of metastases

If the osteosarcoma has spread to other parts of the body, these tumors need to be removed to have a chance at curing the cancer.

Osteosarcoma most often spreads to the <u>lungs</u><sup>4</sup>. If surgery can be done to remove these metastases, it must be planned very carefully. Things to be considered before the operation include:

- The number of tumors
- The location of the tumor(s) (one lung or both lungs)
- The size of the tumor(s)
- How well the tumor(s) responded to chemotherapy
- The person's overall health

adjust to long-term issues such as changes in how they walk or do other tasks, and changes in appearance. Physical, occupational, and other therapies can often help people adjust and cope with these challenges.

# More information about Surgery

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# Chemotherapy and Other Drugs for Osteosarcoma

Chemotherapy (chemo) is the use of drugs to treat cancer. The drugs are usually given into a vein and can reach and destroy cancer cells throughout the body.

Chemo is an important part of the treatment for most people with osteosarcoma (although some patients with low-grade osteosarcoma might not need it). Most osteosarcomas don't appear to have spread beyond the main tumor when they are first found. But in the past, when these cancers were treated with surgery alone, the cancer would often come back in other parts of the body, where it would be very hard to

stays in the vein, while the other end lies just under or outside the skin. This lets the health care team give chemo and other drugs and draw blood samples without having to stick needles into the veins each time. The catheter usually remains in place for several months and can make having chemo less painful. If such a device is used, the health care team will teach you how to care for it to reduce the risk of problems such as infections.

# Side effects of chemo

Chemo drugs can cause <u>side effects</u><sup>3</sup>. Children tend to have less severe side effects from chemo than adults and often recover from side effects more quickly. Because of this, doctors can give them higher doses of chemo to try to kill the tumor.

The side effects of chemo drugs depend on the type, dose, and the length of time they are taken.

#### General side effects of chemo

- Nausea and vomiting
- Loss of appetite
- Diarrhea
- Hair loss
- Mouth sores

Chemo can damage the bone marrow, where new blood cells are made. This can lead to low blood cell counts, which can result in:

- Increased chance of infection (from a shortage of white blood cells)
- Bleeding or bruising after minor cuts or injuries (from a shortage of platelets)
- Fatigue or shortness of breath (from low red blood cell counts)

A major concern with chemo used to treat osteosarcoma is that it can lead to dangerously low white blood cell levels and an increased risk of serious infections. Because of this, drugs called **growth factors** (such as filgrastim, also known as G-CSF) may be given along with the chemo to help the body make new white blood cells as quickly as possible.

Most of the side effects above tend to go away after treatment is finished. Often there are ways to make these side effects less severe. For example, drugs can be given to

help prevent or reduce nausea and vomiting, or to help get blood counts back to normal levels. Be sure to discuss any questions you have about side effects with the cancer care team, and tell them about any side effects so that they can be controlled.

#### Side effects of certain chemo drugs

Some side effects are specific to certain drugs. Many of these side effects are uncommon, but they are possible. Before treatment, ask your cancer care team about the possible side effects of the drugs you or your child will be getting.

- **Ifosfamide** and **cyclophosphamide** can damage the lining of the bladder, which can cause blood in the urine. The chance of this happening can be lowered by giving a drug called **mesna** during chemotherapy, along with plenty of fluids.
- Cisplatin and carboplatincan cause <u>nerve damage (called neuropathy)</u><sup>4</sup> leading to numbness, tingling, or pain in the hands and feet. This often goes away or gets better once treatment is stopped, but it might last a long time in some people. These drugs can sometimes affect hearing, especially of high-pitched sounds. Kidney damage can also occur after treatment. Giving lots of fluid before and after the drug is infused can help prevent this.
- Etoposide can also cause nerve damage. It can also increase the risk of later developing <u>acute myeloid leukemia</u><sup>5</sup>(AML), a cancer of white blood cells. Fortunately, this is not common.
- High-dose methotrexate can damage the white matter of the brain (called leukoencephalopathy) and can also affect the liver and kidneys. Before starting high-dose methotrexate, medicines are given to help protect the kidneys. Methotrexate blood levels may be checked to see how much leucovorin (also called folinic acid) should be given to help limit any damage to normal tissues.
- **Doxorubicin (Adriamycin)** can damage the heart muscle. The risk of this goes up with the total amount of the drug that is given, so doctors are careful to limit the total dose. Your (child's) doctor may order a heart function test (such as an echocardiogram) before and during treatment to see if this drug is affecting the heart. A drug called **dexrazoxane** may be given along with the chemo to help lessen the possible damage.

Some chemo drugs can affect your (child's) **ability to have children (fertility)** later in life. Ask the cancer care team about the possible effects of treatment on fertility, and ask if there are options for <u>preserving fertility</u><sup>6</sup>, such as sperm banking or egg preservation.

The doctors and nurses will watch closely for side effects. Don't hesitate to ask the cancer care team any questions about side effects.

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# Radiation Therapy for Osteosarcoma

Radiation therapy uses high-energy rays or particles to kill cancer cells.

Osteosarcoma cells are not easily killed by radiation, so radiation therapy doesn't play a major role in treating this type of cancer. But sometimes radiation can be useful if the tumor can't be removed completely by surgery. For example, osteosarcoma can start in hip bones or in the bones of the face, particularly the jaw. In these situations, often it's not possible to remove all of the cancer. After as much of the tumor is removed as

possible, radiation is given to try to kill the remaining cancer cells. Chemotherapy is then often given after the radiation.

Radiation can also be used to help slow tumor growth and control symptoms like pain and swelling if surgery is not possible, or if the cancer has <u>come back</u><sup>1</sup>.

## **External beam radiation therapy**

This is the type of radiation therapy most often used to treat osteosarcoma. A machine outside the body focuses high-energy beams on the tumor to kill the cancer cells.

Before treatments start, the radiation team takes careful measurements with imaging tests such as MRI scans to determine the correct angles for aiming the radiation beams and the proper dose of radiation. This planning session is called **simulation**.

Most often, radiation treatments are given 5 days a week for several weeks. Each treatment is much like getting an x-ray, although the dose of radiation is much higher. The treatment is not painful. For each session, you (or your child) will lie on a special table while a machine delivers the radiation from precise angles.

Each treatment lasts only a few minutes, although the setup time – getting into place for treatment – usually takes longer. Young children may be given medicine to make them sleep so they will not move during the treatment.

Newer techniques, such as **intensity modulated radiation therapy (IMRT)**, **conformal proton beam therapy**, and **stereotactic radiosurgery (SRS)**, let doctors aim the radiation at the tumor more precisely while reducing how much nearby healthy tissues get. This may offer a better chance of increasing the success rate and reducing side effects. Many doctors now recommend using these approaches when they are available. (See <u>What's New in Osteosarcoma Research?</u><sup>2</sup>)

#### Possible side effects of radiation therapy

The side effects of external radiation therapy depend on the dose of radiation and where it is aimed.

**Short-term problems** can include effects on skin areas that receive radiation, which can range from mild sunburn-like changes and hair loss to more severe skin reactions. Radiation to the abdomen or pelvis can cause <u>nausea</u><sup>3</sup>, diarrhea, and urinary problems. Talk with your (child's) health care team about the possible side effects because there may be ways to relieve some of them.

In children, radiation therapy can **slow bone growth**. For example, radiation to the bones in one leg might result in it being shorter than the other. Radiation to the facial bones may cause uneven growth, which might affect how a child looks. But if a child is fully or almost fully grown, this is less likely to be an issue.

Depending on where the radiation is aimed, it can also damage other organs:

- Radiation to the chest wall or lungs can affect lung and heart function.
- Radiation to the **jaw area** might affect the salivary glands, which could lead to dry mouth and tooth problems.
- Radiation therapy to the **spine or skull** might affect the nerves in the spinal cord or brain. This could lead to nerve damage, headaches, and trouble thinking, which usually become most serious 1 or 2 years after treatment. Radiation to the spine might cause numbness or weakness in part of the body.
- Radiation to the **pelvis** can damage the bladder or intestines, which can lead to problems with urination or bowel movements. It can also damage reproductive organs, which could affect a child's fertility later in life, so doctors do their best to protect these organs by shielding them from the radiation or moving them out of the way whenever possible.

Another major concern with radiation therapy is that it might cause a new cancer to form in the part of the body that was treated with the radiation. The higher the dose of radiation, the more likely this is to occur, but the overall risk is small and should not keep children who need radiation from getting it.

To lower the risk of serious long-term effects from radiation, doctors try to use o the facial



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- 1. <u>www.cancer.org/treatment/survivorship-during-and-after-treatment/long-term-health-concerns/recurrence.html</u>
- 2. www.cancer.org/cancer/osteosarcoma/about/new-research.html
- 3. <u>www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/nausea-and-vomiting.html</u>
- 4. <u>www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/pain.html</u>
- 5. <u>www.cancer.org/treatment/understanding-your-diagnosis/advanced-cancer/bone-</u> <u>metastases.html</u>
- 6. <u>www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/low-blood-counts.html</u>
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# Treatment Based on the Extent of the Osteosarcoma

Treatment for osteosarcoma depends on several factors, including the location, <u>extent</u>, <u>and grade</u><sup>1</sup> of the cancer, if doctors think it can be removed (resected) completely, and on a person's age and overall health.

## Localized, resectable osteosarcoma

These cancers have not been found to have spread to other parts of the body, and all of the visible tumor can be removed (resected) by surgery.

**High grade:** Most osteosarcomas are high grade, meaning they will probably grow and spread quickly if not treated. The usual treatment for these cancers is as follows:

- Biopsy to establish the diagnosis
- Chemotherapy (chemo), usually for about 10 weeks
- Surgery to remove the tumor, preferably by the same surgeon who did the biopsy. If cancer is found at the edge of the surgery specimen (meaning some cancer might have been left behind), a second surgery might be done to try to remove any remaining cancer. Radiation therapy might be given to the area as well. of the cancer, ifi More chemo (for up to a year). If the initial chemo killed most of the cancer cells, the same drugs are often given again after surgery. If the initial chemo didn't work

#### Localized, non-resectable osteosarcoma

These cancers have not been found to have spread to other parts of the body, but they can't be removed (resected) completely by surgery. For example, they may be too large or too close to vital structures in the body to be resected completely.

As with other osteosarcomas, a biopsy is needed first to establish the diagnosis.

Chemotherapy is usually the first treatment for these cancers. If the tumor shrinks enough to become resectable, it is then removed with surgery. This is followed by more chemotherapy for up to a year.

If the tumor still can't be removed completely after chemotherapy, radiation therapy can often be used to try to keep the tumor under control and to help relieve symptoms. More chemo might be another option, either instead of or after radiation therapy. If the first chemo regimen didn't work very well, different chemo drugs might be tried.

Because these tumors can be hard to treat, a <u>clinical trial</u><sup>2</sup> of newer treatments may be a good option in many cases.

#### Metastatic osteosarcoma

These cancers have already spread to distant parts of the body when they are diagnosed. Most often they have spread to the <u>lungs</u><sup>3</sup>. As with other osteosarcomas, a biopsy is needed first to establish the diagnosis.

Chemotherapy is usually the first treatment for these cancers. If all of the tumors are thought to be resectable after chemotherapy, they are removed with surgery, sometimes in more than one operation. This is followed by more chemo for up to a year.

If some of the tumors remain unresectable after chemo, radiation therapy can often be used to try to keep them under control and to help relieve symptoms. More chemo might be another option, either instead of or after radiation therapy. If the first chemo regimen didn't work very well, different chemo drugs might be tried.

Newer targeted therapy drugs such as regorafenib (Stivarga), sorafenib (Nexavar), or cabozantinib (Cabometyx) might also be an option at some point as well, although more research will be needed to see how effective these drugs are (see <u>What's New in</u> <u>Osteosarcoma Research?</u><sup>4</sup>).

Because these tumors can be hard to treat, <u>clinical trials</u><sup>5</sup> of newer treatments may be a

good option in many cases.

#### **Recurrent osteosarcoma**

Recurrent cancer means that the cancer has come back after treatment. It may come back locally (near where the first tumor was) or in other parts of the body. Most of the time, if osteosarcoma recurs it will be in the lungs.

If possible, surgery to remove the tumor(s) is an important part of treatment, as it offers the best chance for long-term survival. If the cancer recurs at the original site on an arm or leg after limb-sparing surgery, amputation of the limb may be recommended.

Chemotherapy is often part of the treatment for recurrent cancers as well. If the cancer is not resectable, chemo might be used to try to shrink the tumor(s), which might then allow surgery to be done. If the cancer is resectable, chemo might be given after surgery. For more advanced cancers, chemo might be used to try to help relieve symptoms.

Radiation therapy might be part of treatment as well. It can sometimes help keep tumor growth in check and help relieve symptoms.

If the cancer is still growing, newer targeted therapy drugs such as regorafenib (Stivarga), sorafenib (Nexavar), or cabozantinib (Cabometyx) might be an option at some point as well, although more research is needed to see how effective these drugs are (see <u>What's New in Osteosarcoma Research?</u><sup>6</sup>).

Because these tumors can be hard to treat, <u>clinical trials</u><sup>7</sup> of newer treatments may be a good option.

# Hyperlinks

- 1. www.cancer.org/cancer/osteosarcoma/detection-diagnosis-staging/staging.html
- 2. <u>www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html</u>
- 3. <u>www.cancer.org/treatment/understanding-your-diagnosis/advanced-cancer/lung-</u> <u>metastases.html</u>
- 4. <u>www.cancer.org/cancer/osteosarcoma/about/new-research.html</u>
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