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Treating Ewing Tumors

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

How are Ewing tumors treated?

The main goals of treatment for Ewing tumors are:

- To try to cure the patient
- To keep as much function in affected parts of the body as possible
- To limit the long-term complications of treatment as much as possible

Common types of treatment for Ewing tumors include:

- [Chemotherapy for Ewing Tumors](#)
- [Surgery for Ewing Tumors](#)
- [Radiation Therapy for Ewing Tumors](#)
- [High-dose Chemotherapy and Stem Cell Transplant for Ewing Tumors](#)

Common treatment approaches

Treatment will depend on the stage of the cancer and other factors.

Chemotherapy is almost always the first treatment. Localized therapy (surgery and/or radiation therapy) is next, often followed by more chemotherapy. A stem cell transplant might be an option for some patients with Ewing tumors that are unlikely to be cured with other treatments.

- [Treatment of Ewing Tumors by Stage](#)

Who treats Ewing tumors?

Treating Ewing tumors requires a team approach that includes different types of specialists. For children and teens, treatment is best done at a children's cancer center. For adults with Ewing tumors, treatment is typically done 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 **pathologist** (a doctor specializing in lab tests to diagnose and classify diseases)
- A **physiatrist** (a doctor who directs a person's 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, physical therapists and other rehabilitation specialists, and other health professionals.

- [Health Professionals Associated With Cancer Care¹](#)
- [How to Find the Best Cancer Treatment for Your Child²](#)
- [Navigating the Health Care System When Your Child Has Cancer³](#)

Making treatment decisions

The treatment for Ewing tumors 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.

The treatment team will also help you take care of side effects and can help you work closely with nutritionists, psychologists, social workers, and other professionals to understand and deal with medical problems, stress, and other issues related to the 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 to know that the health professionals who treat children with Ewing Tumors are using the experience and knowledge gained from many decades of studying the treatment of this disease.

If time allows, getting a second opinion from another doctor experienced with Ewing tumors 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.

- [Questions to Ask the Doctor About Ewing Tumors](#)⁴
- [How to Talk to Your Child's Cancer Care Team](#)⁵
- [Seeking a Second Opinion](#)⁶

Thinking about taking part in a clinical trial

Today, most children and teens with cancer are treated at specialized children's cancer centers. These centers offer the most up-to-date-treatment by conducting clinical trials (studies of promising new therapies). Children's cancer centers often conduct many clinical trials at any one time, and in fact most children treated at these centers take part in a clinical trial as part of their treatment.

Adults with cancer also typically have the option to participate in a clinical trial. Clinical trials are one way to get state-of-the art cancer treatment. In some cases they may be the only way to get access to newer treatments. They are also the best way for doctors to learn better methods to treat cancer. Still, they might not be right for everyone. Talk to your treatment team to learn about possible clinical trials, and ask about the pros and cons of enrolling in one of them.

If you would like to learn more about clinical trials, start by asking the treatment team if your clinic or hospital conducts clinical trials.

- [Clinical Trials](#)⁷

Considering complementary and alternative methods

You may hear about alternative or complementary methods that the doctor hasn't mentioned. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used *along with* your regular

medical care. Alternative treatments are used *instead of* standard medical treatment. Although some of these methods might be helpful in relieving symptoms or helping people feel better, many have not been proven to work. Some might even be harmful.

Be sure to talk to your cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision.

- [Complementary and Alternative Medicine](#)⁸

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](#)⁹

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](#)¹⁰
- [Find Support Programs and Services in Your Area](#)¹¹

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 him

or her questions about your treatment options.

Chemotherapy for Ewing Tumors

Chemotherapy (chemo) is the use of anti-cancer drugs delivered through a vein (IV) or, rarely, by mouth in the form of pills. These drugs enter the bloodstream and affect cancer cells in all parts of the body, which makes this treatment useful for cancers that are likely to have spread.

Chemo is an important part of treatment for almost all patients with Ewing tumors. It is typically the first treatment given, followed by [surgery](#)¹ and/or [radiation therapy](#)². Then more chemo is often given after the surgery and/or radiation.

As noted in [Ewing Tumor Stages](#)³, even patients with localized Ewing tumors, who have no obvious cancer spread in bone marrow biopsy samples or on imaging tests, are likely to have areas of cancer spread that are too small to be found with these tests. If these patients do not get chemotherapy, these small metastases would eventually develop into larger tumors.

How is chemo given?

Doctors give chemo in cycles, with a period of treatment (often a few days in a row) followed by a rest period to give the body time to recover. A combination of several chemo drugs is used to treat patients with Ewing tumors.

In the United States, the most common regimen is known as **VDC/IE** (or **VAC/IE**). It alternates between 2 combinations of drugs given every 2 to 3 weeks:

- The first set of drugs includes vincristine, doxorubicin (Adriamycin), and cyclophosphamide.
- Once the patient recovers, a combination of ifosfamide and etoposide is given.

Some doctors may use slightly different combinations of drugs.

Most patients will get chemo for at least 9 weeks before surgery or radiation, and then will get more chemo afterward as well. Usually a total of about 14 to 15 cycles of chemo

are given, which can take from about 6 months to close to a year to complete, depending on the schedule. If the tumor has spread, these same drugs may be given at higher doses.

Soon after the Ewing tumor is diagnosed (but before starting chemo), the doctor may suggest surgery to put a catheter (a thin, soft tube) into a large vein in the patient's chest. This is sometimes called a [venous access device \(VAD\) or central venous catheter \(CVC\)](#)⁴. One end of the catheter stays in the vein, while the other end lies just under or outside the skin. This lets the healthcare team give chemo and other drugs and to draw blood samples without having to stick needles into the veins each time. The catheter usually stays in place for several months, and can make having chemo less painful. If you or your child gets such a device, the health care team will teach you how to care for it to reduce the risk of problems such as infections.

Possible side effects of chemo

Chemo drugs can affect cells in the body other than cancer cells, which can lead to side effects. The side effects depend on the type and doses of drugs, and the length of time they are given.

Children tend to have less severe side effects from chemo than adults and often recover from side effects more quickly. This is why doctors can often give them higher doses of chemo to kill the tumor.

General side effects: Side effects common to many chemo drugs include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue (from having too few red blood cells)

Most of these side effects tend to go away after treatment is finished. There are often ways to lessen them. For example, drugs can be given to help prevent or reduce nausea and vomiting, or to help get blood cell counts back to normal levels. Be sure to discuss any questions you have about side effects with the cancer care team.

Side effects of certain drugs: Along with the effects listed above, certain chemo drugs can have specific side effects:

- **Cyclophosphamide** and **ifosfamide** can damage the bladder, which can cause blood in the urine. The risk of this happening can be lowered by giving the drugs with plenty of fluids and with a drug called *mesna*, which helps protect the bladder.
- **Doxorubicin** can damage the heart. This risk goes up as the total dose of the drug goes up. Doctors try to limit this risk as much as possible by not giving more than the recommended doses of doxorubicin and by checking the heart with a test called an *echocardiogram* during treatment.
- **Vincristine** can damage [nerves](#)⁵. Some patients may notice tingling and numbness, particularly in the hands and feet. This often goes away or gets better once treatment is stopped, but it may last a long time in some people.
- Some chemo drugs can affect your (child's) ability to have children later in life. Talk to the cancer care team about the risks of infertility with treatment, and ask if there are options for preserving fertility, such as sperm banking or ovarian tissue banking.
- Some drugs used to treat Ewing tumors, such as **etoposide**, can increase the risk of developing leukemia later on, although this is not common.

For more on the possible late or long-term side effects of chemo, including infertility and second cancers, see [Living as a Ewing Tumor Survivor](#)⁶.

Tests to check for chemo side effects: Before each treatment, lab tests will be done to be sure the liver, kidney, and bone marrow are working well. If not, chemo may need to be delayed or the doses reduced.

- The **complete blood count (CBC)** includes counts of white blood cells, red blood cells, and blood platelets. Chemo can lower the numbers of these blood cells, so blood counts will be watched closely during and after chemo. The white blood cells and platelets usually reach their lowest point about 2 weeks after chemo is given, though this can occur earlier with high-dose regimens.
- **Blood chemistry tests** measure certain chemicals in the blood that tell doctors how well the liver and the kidneys are working. Some chemo drugs can damage the kidneys and liver.
- If doxorubicin (Adriamycin) is to be given, tests such as an **echocardiogram** (an ultrasound of the heart) may be done to check heart function before and during treatment.

Hyperlinks

1. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/surgery.html>
2. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
3. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/detection-diagnosis-staging/staging.html>
4. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/central-venous-catheters.html>
5. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects/peripheral-neuropathy.html>
6. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/follow-up.html>
7. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html>
8. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects.html>

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see [Chemotherapy](#)⁷.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)⁸.

References

Anderson ME, Randall RL, Springfield DS, Gebhart MC. Chapter 92: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Gebhart MC, DuBois S. Treatment of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/treatment-of-the-ewing-sarcoma-family-of-tumors on March 8, 2018.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2018. Accessed at

www.nccn.org/professionals/physician_gls/pdf/bone.pdf on March 8, 2018.

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Surgery for Ewing Tumors

Surgery is an important part of treatment for most Ewing tumors. Surgery is usually done for two reasons:

- To diagnose the cancer (with a [biopsy](#)¹)
- To remove the tumor(s)

Whenever possible, it's very important that the biopsy and surgical treatment be planned together, and that the same orthopedic surgeon at a cancer center does both the biopsy and the surgery to remove the tumor. The biopsy needs to 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 number of cancer cells are left behind, they might grow and multiply to make a new tumor. To lower the risk of this happening, surgeons remove the tumor plus some of the normal tissue that surrounds it. This is known as *wide excision*.

Using a microscope, a doctor called a pathologist will look at the removed tissue to see if the margins (outer edges) have cancer cells.

- 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 grow back in the place where it started.

Types of surgery for Ewing tumors

Many types of surgery can be used for Ewing tumors. The choice depends on the tumor's size and location, the age of the patient, how likely it is that surgery can remove the tumor with clean margins, and how surgery would change the function of the affected part of the body.

Tumors in some soft tissues and certain bones can be removed without causing major disability or deformity. Other tumors, such as those in the bones of the arms and legs, might not be able to be removed completely without affecting the limb's function.

Although all operations to remove Ewing sarcomas are complex, tumors in the arms or legs are generally not as hard to remove as those in other parts of the body, such as the base of the skull, the chest wall, the spine, or the pelvis (hip bones).

Tumors in the arms or legs

For most tumors in an arm or leg, surgery can remove part or all of the affected bone while leaving the arm or leg basically intact. This is known as **limb-sparing surgery**. The bone that is removed is replaced either with a bone graft (a piece of bone from another part of the body or from another person) or with an internal prosthesis (a rod-shaped device made of metal and other materials that replaces part or all of a bone). Some newer devices combine a graft and a prosthesis.

If the tumor is in the upper part of the leg, the femur (upper leg bone), including the knee, can be removed and replaced with a prosthesis for the bone and knee, which is connected to the lower leg. Tumors in the lower part of the leg are harder to treat this way, because it is harder to remove and reconstruct parts of the lower leg. The humerus (upper arm bone) is also suitable for limb-sparing surgery.

Limb-sparing surgery is a very complex operation. The surgeons who do this type of operation must have special skills and experience. The challenge for the surgeon is to be sure 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 tumor has grown into these structures, they will need to be removed as well. In such cases, [radiation therapy](#)² or **amputation** (removal of part or all of the limb) may sometimes be the best option to treat the tumor.

Using an internal prosthesis in a growing child is especially challenging. In the past, it often required several operations over time to replace the prosthesis with a longer one as the child grew. Newer prostheses have become very sophisticated. Some 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.

Some children may not be able to have limb-sparing surgery because their tumors are in parts of bones that are hard to replace or because the tumors also extend into vital nerves or blood vessels that can't be removed without severely damaging the limb. These children usually get radiation therapy instead of surgery. In rare cases, amputating the affected limb may be the best option.

Tumors in the chest wall or pelvis (hip bones)

For a Ewing tumor in the chest wall, the surgeon often must remove the diseased area and also remove nearby ribs. The ribs might then be replaced with a man-made material. If the tumor has spread to the lungs, the chest can be opened and the lung tumors removed during an operation called a *thoracotomy*. Often these patients also get [radiation therapy](#)³ to the chest.

Pelvic tumors can be hard to treat with surgery, and in many cases radiation therapy may be the preferred treatment. But if the tumor responds well to initial [chemotherapy](#)⁴, surgery (sometimes followed by radiation therapy) may be an option. 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.

Tumors in the spine

Tumors in or right next to the spine can often be hard to remove completely, so [radiation therapy](#)⁵ is sometimes a better option. If surgery is done, radiation is often given afterward to try to kill any remaining tumor cells.

Possible side effects of surgery

Short-term risks and side effects: Surgery to remove a Ewing tumor is often a long and complex operation. Serious short-term side effects are not common, but they can include reactions to anesthesia, excess bleeding, blood clots, and infections. Pain is common after the operation, and strong pain medicines might be needed for a while after surgery as the site heals.

Long-term side effects: The long-term side effects of surgery depend mainly on where the tumor is and what type of operation is done.

Complications of limb-sparing surgery can include bone grafts or prostheses that might break or become loose. This is more likely than with bone surgery done for other

reasons because the [chemo](#)⁶ used before and after surgery can increase the risk of [infection](#)⁷ and affect wound healing. Infections in the area can be very serious because they can be hard to treat, and might require further surgery. Infections are also a concern in people who have had amputations, especially of part of a leg, because the pressure placed on the skin at the site of the amputation can cause the skin to break down over time.

Rehabilitation after surgery

This might be the hardest part of treatment, and can't be described here completely because it will be different for each patient. Whenever possible, patients and parents should meet with a rehabilitation specialist before surgery to learn about their options and what might be required after surgery.

Physical therapy and rehabilitation are very important for patients who have had surgery for Ewing tumors. Following the recommended rehab program offers the best chance for good long-term limb function. Even with proper rehab, people might still have to 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.

Rehab after limb-sparing surgery

Even when only the tumor and part of the bone is removed in a limb-sparing operation, the situation can still be complicated, especially in growing children. Children who have had limb-sparing surgery may need more surgery in the coming years to replace the internal prosthesis with one more suited to their growing body size, and some may eventually need an amputation.

It takes about a year, on average, for patients to learn to walk after limb-sparing surgery on a leg. Physical rehabilitation after limb-sparing surgery is extremely important. If the patient doesn't actively take part in the rehabilitation program, the salvaged arm or leg can become useless.

Rehab after amputation

If a limb is amputated, the patient must learn to adjust to new ways of doing some things, often with the use of a prosthetic limb. This can be particularly hard for growing children if the prosthetic limb needs changing to keep up with their growth. With proper physical therapy, patients are often able to walk on their own about 3 to 6 months after a leg amputation.

Considering your options

Both limb-sparing surgery and amputation can have pros and cons. For example, limb-sparing surgery, although often preferred by patients over amputation, tends to lead to more complications because of its complexity. Growing children who have limb-sparing surgery are also more likely to need further surgery later on.

When researchers have looked at the results of the different surgeries in terms of quality of life, there has been little difference between them. Perhaps the biggest problem has been for teens, who may worry about the social effects of their operation. Emotional issues can be very important, and all patients will need support and encouragement. See [Living as a Ewing Tumor Survivor](#)⁸.

Hyperlinks

1. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/detection-diagnosis-staging/how-diagnosed.html>
2. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
3. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
4. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
5. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
6. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
7. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects/infections/infections-in-people-with-cancer.html>
8. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/follow-up.html>
9. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/treatment-types/surgery.html>
10. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects.html>

More information about Surgery

For more general information about surgery as a treatment for cancer, see [Cancer Surgery](#)⁹.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)¹⁰.

References

Anderson ME, Randall RL, Springfield DS, Gebhart MC. Chapter 92: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Gebhart MC, DuBois S. Treatment of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/treatment-of-the-ewing-sarcoma-family-of-tumors on March 8, 2018.

Hornicek FJ. Bone sarcomas: Preoperative evaluation, histologic classification, and principles of surgical management. UpToDate. Accessed at www.uptodate.com/contents/bone-sarcomas-preoperative-evaluation-histologic-classification-and-principles-of-surgical-management on March 9, 2018.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2018. Accessed at www.nccn.org/professionals/physician_gls/pdf/bone.pdf on March 8, 2018.

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Radiation Therapy for Ewing Tumors

Radiation therapy focuses high-energy beams at the tumor from a machine outside the body to kill the cancer cells.

Ewing tumors are very sensitive to radiation, so radiation therapy can sometimes be helpful in treating them. It may be used with [surgery](#)¹, or it may be used instead of surgery, especially if it would be hard to remove the entire tumor. In either case, [chemotherapy](#)² is usually given before, during, and afterward.

How radiation therapy is done

This type of treatment is given by a doctor called a *radiation oncologist*. Before treatments start, the radiation team takes careful measurements with imaging tests such as [MRI scans](#)³ to determine the correct angles for aiming the beams and the proper dose of radiation. This planning session is called *simulation*. Patients may also be fitted with a plastic mold resembling a body cast to keep them in the same position each time so that the radiation can be aimed more accurately.

Most often, radiation treatments are given 5 days a week for several weeks. Each treatment is much like getting an x-ray, but the dose of radiation is much higher. The treatment is not painful. For each session, the patient lies on a special table while a machine delivers the radiation from precise angles.

Each treatment lasts only a few minutes, but the setup time – getting the patient into place for treatment – usually takes longer. Some younger children may be given medicine before each treatment to make them sleep so they won't move during treatment.

Types of radiation therapy

Modern radiation therapy techniques let doctors focus the radiation more precisely than in the past. These include:

Three-dimensional conformal radiation therapy (3D-CRT): 3D-CRT uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor. Several radiation beams are then shaped and aimed at the tumor from different directions. Each beam alone is fairly weak, which makes it less likely to damage normal body tissues, but the beams converge at the tumor to give a higher dose of radiation there.

Intensity modulated radiation therapy (IMRT): IMRT is an advanced form of 3D therapy that can be especially useful for tumors near the spine. Along with shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams is adjusted to limit the dose reaching the most sensitive normal tissues. This lets the doctor deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT, especially for tumors in hard-to-treat areas such as the spine or pelvis (hip bones).

Conformal proton beam radiation therapy: Proton beam therapy is another type of 3D therapy. But instead of using x-rays, it focuses proton beams on the tumor. Unlike x-

rays, which release energy both before and after they hit their target, protons cause little damage to tissues they pass through and then release their energy after traveling a certain distance. Doctors can use this property to deliver more radiation to the tumor and do less damage to nearby normal tissues.

This approach may be helpful for hard-to-treat tumors, such as those on the spine, skull, or pelvic bones. The machines needed to make protons are expensive, and there are a limited number of them being used in the United States at this time.

Possible side effects of radiation therapy

Because of the possible side effects of radiation therapy (especially in growing children), surgery is often preferred if it is possible. But improvements in the way radiation therapy is given now allow children with Ewing tumors to be treated with lower doses than were used in the past, helping to reduce some of these side effects.

The side effects of radiation therapy depend mainly on the dose of radiation and where it is aimed. Some effects may be short term, while others may have a longer lasting impact.

Short-term problems include effects on the skin in areas that receive radiation, which can range from mild sunburn-like changes and hair loss to more severe skin reactions. Radiation might also lower blood cell counts. Radiation to the abdomen or pelvis can cause nausea, diarrhea, and urinary problems.

Long-term side effects can be more serious, especially in growing children, so doctors try to limit them as much as possible.

A serious effect of radiation therapy in children is **slowed bone growth**, especially in younger children. For example, radiation to the bones in one leg may result in it being much shorter than the other. Radiation of 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 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 [fertility](#)⁴ 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.

- Side effects of **radiation therapy to the spinal cord or skull** may include nerve damage, headaches, and trouble thinking, which usually become most serious 1 or 2 years after treatment. Fortunately, Ewing tumors rarely spread to the brain, but they can sometimes extend into the brain from nearby bones of the skull.

Another major concern with radiation therapy is that it might cause a [new cancer](#)⁵ to form in the part of the body that was treated with the radiation. This is most often a different type of bone cancer called [osteosarcoma](#)⁶. This small risk should not keep children who need radiation from getting it. Still, it's important to continue follow-up visits with your child's doctor so that if problems come up they can be found and treated as early as possible.

Hyperlinks

1. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/surgery.html>
2. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
3. <https://www.cancer.org/content/cancer/en/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html>
4. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects/fertility-and-sexual-side-effects.html>
5. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects/second-cancers-in-adults.html>
6. <https://www.cancer.org/content/cancer/en/cancer/osteosarcoma.html>
7. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/treatment-types/radiation.html>
8. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects.html>

More information about radiation therapy

To learn more about how radiation is used to treat cancer, see [Radiation Therapy](#)⁷.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)⁸.

References

Anderson ME, Randall RL, Springfield DS, Gebhart MC. Chapter 92: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Gebhart MC, DuBois S. Treatment of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/treatment-of-the-ewing-sarcoma-family-of-tumors on March 8, 2018.

DeLaney TF. Radiation therapy for Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/radiation-therapy-for-ewing-sarcoma-family-of-tumors on March 8, 2018.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2018. Accessed at www.nccn.org/professionals/physician_gls/pdf/bone.pdf on March 8, 2018.

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High-dose Chemotherapy and Stem Cell Transplant for Ewing Tumors

This type of treatment is being studied for Ewing tumors that are hard to cure with other treatments, such as tumors that have spread (metastasized) to other parts of the body or that have come back after standard treatment. So far, it's not clear if a stem cell transplant is better than other treatments (such as standard chemotherapy), so many doctors recommend it be done only as part of a [clinical trial](#)¹.

What is a stem cell transplant?

The doses of [chemotherapy](#)² (chemo) drugs that can be given safely are normally limited by the side effects these drugs can cause. One of the most serious is damage to the stem cells in bone marrow, which is where new blood cells are made. Even though higher doses of chemo might be more effective in treating Ewing tumors, they can't be given because they would severely damage these bone marrow cells, leading to life-

threatening shortages of blood cells.

To try to get around this problem, high-dose chemo (sometimes along with radiation therapy) can be followed by a stem cell transplant to “rescue” the bone marrow, giving the person new blood stem cells to replace those that were destroyed.

In the past, this type of treatment was often called a *bone marrow transplant*.

If a stem cell transplant is considered as part of the initial treatment plan for a Ewing tumor, the person first gets standard doses of chemo, then local treatment of the tumor ([surgery](#)³ and/or [radiation therapy](#)⁴), followed by high-dose chemo and a stem cell transplant.

How a stem cell transplant is done

The first step in a stem cell transplant is done before the treatment with high-dose chemo. The patient's own blood-producing stem cells are collected (harvested) to use later. This type of transplant, where the stem cells are taken from the patient, is known as an **autologous transplant**. (Another type of stem cell transplant, called **allogeneic transplant**, uses stem cells from a donor. This type is not used often for treating Ewing tumors.)

The stem cells are usually collected from the bloodstream using a procedure similar to a blood donation. But instead of going into a collecting bag, the blood goes into a special machine that filters out the stem cells and returns the other parts of the blood to the person's body. The stem cells are then frozen until the transplant. This may need to be done more than once.

Once the stem cells have been frozen and stored, the person gets high-dose chemo, sometimes along with radiation therapy. When the treatment is finished, the patient's stem cells are thawed and returned to the body in a blood transfusion. The stem cells travel through the bloodstream and settle in the bone marrow. Over the next few weeks, they start to make new, healthy blood cells.

Side effects of stem cell transplants

A stem cell transplant is a complex treatment that can cause serious or even life-threatening side effects. If the doctors think a person might benefit from a transplant, it should be done at a cancer center where the staff has experience in doing the procedure and managing the recovery period.

Some side effects of a stem cell transplant might last a long time, or might not show up until years after the transplant, which is an especially important concern in children and teens. If a stem cell transplant is recommended for your child, be sure to talk to the cancer care team before the transplant to learn about possible long-term effects your child might have.

Hyperlinks

1. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/clinical-trials.html>
2. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
3. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/surgery.html>
4. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
5. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/treatment-types/stem-cell-transplant.html>
6. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/physical-side-effects.html>

More information about stem cell transplant

To learn more about stem cell transplants, including how they are done and their potential side effects, see [Stem Cell Transplant for Cancer](#)⁵.

For more general information about side effects and how to manage them, see [Managing Cancer-related Side Effects](#)⁶.

References

Anderson ME, Randall RL, Springfield DS, Gebhart MC. Chapter 92: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Gebhart MC, DuBois S. Treatment of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/treatment-of-the-ewing-sarcoma-family-of-tumors on March 8, 2018.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2018. Accessed at www.nccn.org/professionals/physician_gls/pdf/bone.pdf on March 8, 2018.

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Treatment of Ewing Tumors by Stage

Treatment of a Ewing tumor is based mainly on where it is in the body and [how far it has spread](#)¹ when it's first found.

Localized Ewing tumors

A localized Ewing tumor is one that still appears to be confined to the area where it started (and maybe also nearby tissues such as muscle or tendons), based on [imaging test and biopsy results](#)². But even people with localized Ewing tumors often still have cancer spread to other parts of the body that is too small to be seen with imaging tests. If these people do not get chemotherapy, these small areas of cancer cells would eventually become larger tumors. This is why chemotherapy, which can reach all parts of the body, is an important part of treatment for localized Ewing tumors.

Once the Ewing tumor has been diagnosed and staged, the first treatment is [chemotherapy](#)³. It's called *neoadjuvant* chemotherapy because it's given before any [surgery](#)⁴ or [radiation therapy](#)⁵. In the United States, treatment is usually a regimen known as VDC/IE (or VAC/IE), which is a combination of vincristine, doxorubicin (Adriamycin), and cyclophosphamide, alternated with ifosfamide and etoposide, although other combinations of the same drugs are also effective.

After at least 9 weeks of chemotherapy, imaging tests such as CT, MRI, PET, or bone scans are done to see if the tumor is shrinking and can be surgically removed.

If so, surgery is done at this point. If cancer cells are found at or near the edges of the surgery specimen (meaning cancer cells may have been left behind), radiation therapy and chemotherapy (for several months) are used. If there are no cancer cells at or near the edges of the surgery specimen, chemotherapy can be used without radiation therapy.

If surgery is not an option after the initial chemotherapy (because of the tumor location or some other reason), but the tumor is not growing, radiation therapy (along with chemotherapy) is usually the next treatment given. In some cases this might shrink the tumor enough so that surgery can then be done. This would then be followed by more chemotherapy, possibly with more radiation as well. In other cases where surgery is still not an option, radiation therapy and chemotherapy are the main treatments.

If the Ewing tumor continues to grow despite the initial chemotherapy, a second type of chemotherapy (using different drugs) may be tried. Surgery or radiation therapy may also be tried to help keep the tumor under control. This may be followed by more chemotherapy.

Metastatic Ewing tumors

Patients who clearly have metastatic disease when they are first diagnosed are harder to treat than patients with localized disease. The outlook tends to be better when the cancer has only spread to the lungs, as opposed to when the cancer has spread to other bones or to the bone marrow.

Treating metastatic disease is similar in many ways to treating localized disease. [Chemotherapy](#)⁶ is the first treatment, but it often requires a more intense regimen than would be used if the cancer was localized. After a few months, tests such as CT or MRI scans, bone or PET scans, and/or bone marrow biopsies are done to see how the cancer has responded to treatment.

If the cancer has only spread to a few small areas, the main (primary) tumor and all known areas of metastases may be removed with [surgery](#)⁷ at this point. Other options, such as surgery plus radiation therapy (before and/or after surgery) or [radiation therapy](#)⁸ alone to all known metastatic sites, might also be tried. During and after these treatments, chemotherapy is given for several months as well.

Doctors at several cancer centers are now studying giving very intensive chemotherapy followed by a [stem cell transplant](#)⁹ to try to improve the outcome for these patients.

Because these tumors can be hard to treat, [clinical trials](#)¹⁰ of newer treatments may be a good option in many cases.

Ewing tumors that recur (come back) after treatment

Recurrence of Ewing tumors after treatment is less likely now than in the past, but it can happen. If the tumor does come back, treatment depends on a number of factors,

including:

- The size and location of the tumor
- Whether it has spread to different parts of the body
- What types of treatment were used before
- How long it has been since treatment

[Chemotherapy](#)¹¹, [surgery](#)¹², [radiation therapy](#)¹³, or some combination of these may be used to treat recurrent tumors, depending on the situation. Doctors are also studying the use of [high-dose chemotherapy followed by a stem cell transplant](#)¹⁴, as well as the use of [targeted drugs and immune therapies](#)¹⁵, but it is not yet clear how useful these are. These tumors can be hard to treat, so [clinical trials](#)¹⁶ of newer treatments may be a good option.

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 doctors. Your doctors may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask them questions about your treatment options.

Hyperlinks

1. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/detection-diagnosis-staging/staging.html>
2. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/detection-diagnosis-staging/how-diagnosed.html>
3. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
4. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/surgery.html>
5. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
6. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
7. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/surgery.html>
8. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>

9. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/high-dose-chemo-stem-cell.html>
10. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/clinical-trials.html>
11. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/chemotherapy.html>
12. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/surgery.html>
13. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/treating/radiation-therapy.html>
14. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/about/new-research.html>
15. <https://www.cancer.org/content/cancer/en/cancer/ewing-tumor/about/new-research.html>
16. <https://www.cancer.org/content/cancer/en/treatment/treatments-and-side-effects/clinical-trials.html>

References

Anderson ME, Randall RL, Springfield DS, Gebhart MC. Chapter 92: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Gebhart MC, DuBois S. Treatment of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/treatment-of-the-ewing-sarcoma-family-of-tumors on March 8, 2018.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2018. Accessed at www.nccn.org/professionals/physician_gls/pdf/bone.pdf on March 8, 2018.

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