Treating Ewing Tumors

If you or your child has been diagnosed with a Ewing tumor (Ewing sarcoma), the cancer care team will discuss treatment 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

The main 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 (extent) 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 **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 Care
- How to Find the Best Cancer Treatment for Your Child
- Navigating the Health Care System When Your Child Has Cancer

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.

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.

Once treatment starts, members of the treatment team can help you deal with side effects, stress, financial aid, 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

Be sure to talk to your cancer care team about any method you are thinking about

Chemotherapy for Ewing Tumors

Chemotherapy (chemo) is the use of drugs to treat cancer. The drugs are usually given into a vein (IV) and can affect cancer cells in all parts of the body, which makes this treatment useful for cancers that are likely to have spread.

When is chemo used for Ewing tumors?

Chemo is an important part of treatment for almost all patients with Ewing tumors (Ewing sarcomas). It is typically the first treatment given, followed by surgery and/or radiation therapy. More chemo is often given after surgery and/or radiation.

As noted in Ewing Tumor Stages[1], even patients with localized Ewing tumors, who have no obvious cancer spread shown on imaging tests or in bone marrow biopsy samples, are likely to have very small areas of cancer spread (micrometastases). If chemotherapy isn't given, these small metastases eventually would 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 Ewing tumors.

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

- The first drug combination includes **vincristine**, **doxorubicin (Adriamycin)**, and **cyclophosphamide**.
- The second set of drugs includes a combination of **ifosfamide** and **etoposide**.

Some doctors may use slightly different drug combinations. These often include many of the same chemo drugs listed above, although other drugs might be used as well (especially if the cancer comes back after treatment).

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 to other parts of the body, 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)[2]. One end of the catheter stays in the vein, while the other end lies just under or outside the skin. The catheter usually stays in place for several months, letting the health care team give drugs and draw blood samples without having to stick needles into the veins each time. 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.

Side effects of chemo

When chemo drugs affect cells in the body other than cancer cells, it 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 of chemo

Common side effects of many chemo drugs include:

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

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 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 chemo 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 (Adriamycin)** can damage the heart. This risk goes up with the total dose of the drug, so doctors are careful to limit the total dose. The doctor might 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.
- **Vincristine** can cause nerve damage (neuropathy)[3]. 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. **Etoposide** can increase the risk of developing leukemia later on, although this is not common.

how well the liver and the kidneys are working. Some chemo drugs can damage these organs.
- 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.

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see Chemotherapy[6].

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects[7].

Hyperlinks

1. www.cancer.org/cancer/ewing-tumor/detection-diagnosis-staging/staging.html
2. www.cancer.org/treatment/treatments-and-side-effects/planning-managing/tubes-lines-ports-catheters.html
3. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/peripheral-neuropathy.html
4. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/fertility-and-sexual-side-effects/preserving-fertility-in-children-and-teens-with-cancer.html
5. www.cancer.org/cancer/ewing-tumor/follow-up.html
6. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html
7. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html

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Surgery for Ewing Tumors

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

Types of surgery for Ewing tumors

Many types of surgery can be used for Ewing tumors. The choice depends on several factors, including:

- The tumor's size and location
- The patient's age and overall health
- How likely it is that surgery can remove the tumor with clean margins
- 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 might not be able to be removed completely without affecting the function or appearance of that part of the body.

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** (or **limb-salvage 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, part of 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.

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 and can often 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 patients 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 patients usually get radiation therapy instead of surgery.

In rare cases, **amputation** (removal of part or all of the limb) may be the best option, especially if the cancer comes back in the place where it started, and radiation therapy has already been used.

For an amputation, 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 prosthesis remaining outside the skin. This can then be attached to an external prosthesis.

Tumors in the chest wall

For a Ewing tumor in the chest wall, the surgeon often must remove the diseased area and also remove nearby ribs, which 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**. Radiation therapy is often given to the chest as well.

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given afterward to try to kill any remaining tumor cells.

Possible side effects of surgery for Ewing tumors

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 possible breaking or loosening of bone grafts or prostheses. This is more likely than with bone surgery done for other reasons because the chemo used before and after surgery can increase the risk of infection 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 it 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 might 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[2].)

diagnosed.html
2. www.cancer.org/cancer/ewing-tumor/follow-up.html
3. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/surgery.html
4. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html

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case, chemotherapy 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[1] to determine the correct angles for aiming the beams and the right 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 techniques 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 with complex shapes or tumors near important structures, such as 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.

3. www.cancer.org/treatment/survivorship-during-and-after-treatment/long-term-health-concerns/second-cancers-in-adults.html
4. www.cancer.org/cancer/osteosarcoma.html
5. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation.html
6. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html

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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.

The main side effects from a stem cell transplant are from the chemotherapy. Because high doses of chemo are used, some of these side effects might be more severe than with standard doses of chemo.

Some side effects of a stem cell transplant might last a long time, or they might not show up until years after the transplant, which is a special 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.

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[2].

For more general information about side effects and how to manage them, see Managing Cancer-related Side Effects[3].

Hyperlinks

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