Treating Ewing Tumors

Treatment overview for Ewing tumors

Once a Ewing tumor has been found and staged, the cancer care team will talk with you about treatment options. It’s important to be sure you understand your child’s options as well as their possible side effects to help make the decision that’s the best fit for your child. If there is anything you don’t understand, ask to have it explained. You can find some good questions to ask in “What should you ask the doctor about Ewing tumors?”

The main goals of treatment of 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

For children and teens, a team approach is recommended that includes the child’s pediatrician as well as children’s cancer specialists. Treatment for children and teens is best done at a children’s cancer center. For adults with Ewing tumors, the treatment team typically includes the patient’s primary care doctor, as well as specialists 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 adults and children, the team will also include other doctors, physician assistants (PAs), nurse practitioners (NPs), nurses, psychologists, social workers, physical therapists and other rehabilitation specialists, and other health professionals. Going
through cancer treatment often means meeting lots of specialists and learning about parts of the medical system you probably haven’t been exposed to before. For more information, see *Children Diagnosed With Cancer: Understanding the Health Care System.*

Before treatment, the doctors and other members of the team will help you, as a parent, understand the tests that will need to be done. The team’s social worker will also counsel you about the problems you and your child might have during and after treatments such as surgery, and might be able to help you find housing and financial aid if needed.

The types of treatment that can be used in Ewing tumors include:

- **Chemotherapy**
- **Surgery**
- **Radiation therapy**
- **High-dose chemotherapy followed by a stem cell transplant**

*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.

Your child’s treatment will depend on the stage of the cancer and other factors. See “*Treatment of Ewing tumors by stage*” for information about the most common approaches.

**Thinking about taking part in a clinical trial**

Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. 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 are not right for everyone.

Most patients with Ewing tumors are treated in clinical trials according to national treatment guidelines called *protocols.* In the United States, some of the most successful protocols have been those of the Children’s Oncology Group and its forerunners. Studies from similar groups in Europe have also produced very important information. Most advances in the treatment of Ewing tumors have come from the results of these clinical trials.
If you would like to learn more about clinical trials that might be right for your child, start by asking your child’s doctor if your clinic or hospital conducts clinical trials. See Clinical Trials to learn more.

**Considering complementary and alternative methods**

You may hear about alternative or complementary methods that your doctor hasn’t mentioned to treat your child’s cancer or relieve symptoms. 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 regular medical care. Alternative treatments are used instead of a doctor’s medical treatment. Although some of these methods might be helpful in relieving symptoms or helping you feel better, many have not been proven to work. Some might even be dangerous.

Be sure to talk to your child’s 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. See the Complementary and Alternative Medicine section to learn more.

**Help getting through cancer treatment**

Your child’s 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 are an important part of your child’s care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help.

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

*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 just about all patients with Ewing tumors. It is typically the first treatment given, followed by surgery and/or radiation therapy. More chemo is often given after the surgery and/or radiation is done.

As noted in the section “How are Ewing tumors staged?”, even patients with localized Ewing tumors, who have no obvious cancer spread in bone marrow 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.

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 alternates between 2 combinations of drugs given every 2 to 3 weeks (VAdriaC alternating with IE). The first set of drugs includes vincristine, doxorubicin (Adriamycin), and cyclophosphamide. After the patient recovers from the effects of these drugs, another combination of drugs, ifosfamide and etoposide, is given. Some doctors may use slightly different combinations of drugs.

Chemotherapy is given for at least 12 weeks before surgery or radiation and is then given 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 putting 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). The catheter is inserted surgically while the patient is under general anesthesia (in a deep sleep). 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 such a device is used in your child, 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 chemotherapy**
Chemo drugs attack cells that are dividing quickly, which is why they work against cancer cells. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemo, which can lead to side effects.

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.

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

**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**. 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 be long lasting in some people.

Some chemo drugs can affect your (child’s) ability to have children later in life. Talk to your (or your child’s) 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 later developing a cancer of white blood cells known as acute myeloid leukemia. Fortunately, this doesn’t happen often.

For more on the possible late or long-term side effects of chemo, including infertility and second cancers, see “What happens after treatment for Ewing tumors?”

**Tests to check for chemo side effects:** Before each treatment, your (child’s) doctor will check lab test results 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.

For more on chemotherapy, see the Chemotherapy section of our website.

- References
See all references for Ewing Family of Tumors

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Surgery for Ewing Tumors

Surgery is an important part of treatment for virtually all Ewing tumors. It includes:

- The biopsy to diagnose the cancer
- The surgical treatment 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 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 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 where it started.

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, 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, often can’t 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 (a limb-sparing operation). The bone that is
removed is replaced either with a bone graft (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).

If the tumor is in the upper part of the leg, the femur (upper leg bone), including the knee, can be removed. It is 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 or amputation 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 are then replaced with a man-made material. If the child’s tumor has spread to the lungs, the chest can be opened and the lung tumors removed during an operation called a *thoracotomy*. Often these children also get radiation therapy 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, 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.

**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 the patient might need strong pain medicines 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 become loose or broken. This is more likely than with surgery done for other reasons because **chemo** used before and after surgery can increase the risk of **infection** and affect wound healing. 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. Patients and parents should meet with a rehabilitation specialist to understand all of their options.

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-salvage surgery is extremely important. If the patient doesn’t actively take part in the rehabilitation program, the salvaged arm or leg can become useless.

If a limb is amputated, the patient must learn to live with and use 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 3 to 6 months after a leg amputation.

Both limb-sparing surgery and amputation can have pros and cons. For example, limb-sparing surgery, although often more acceptable than 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 down the road.

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 “Social, emotional, and other issues in treating Ewing tumors”).

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.

For more on surgery as a treatment for cancer, see A Guide to Cancer Surgery.

• References

See all references for Ewing Family of Tumors

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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. In people with Ewing tumors, radiation therapy may be used with surgery, or it may be used instead of surgery, especially if it would be hard to remove the entire tumor. In either case, chemotherapy is usually given before, during, and afterward.
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 to determine the correct angles for aiming the beams and the proper dose of radiation. Your child may be fitted with a plastic mold resembling a body cast to keep him or her 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, your child will lie 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 your child 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.

Some newer techniques let doctors focus the radiation more precisely:

**Three-dimensional conformal radiation therapy (3D-CRT):** Three-dimensional 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 related to 3D-CRT. 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 or
pelvic bones. The machines needed to make protons are expensive, and there are only a handful 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 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 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 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 on bone growth. In younger children, some bones will not grow well after radiation. 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 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 may cause a new cancer 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. The higher the dose of radiation, the more likely this is to occur. This small risk should not keep children who need radiation from getting treatment. 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.

For more on radiation therapy, see the Radiation Therapy section of our website or Understanding Radiation Therapy: A Guide for Patients and Families.

- References

See all references for Ewing Family of Tumors

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High-dose Chemotherapy and Stem Cell Transplant for Ewing Tumors

This type of treatment is being studied for patients with Ewing tumors that are hard to cure with other treatments, such as those with metastatic disease or with Ewing tumors that come back after the standard treatment.

The doses of chemotherapy (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 bone marrow, which is where new blood cells are made. Even though higher doses of these drugs might be more effective in treating Ewing tumors, they can’t be given because they would severely damage bone marrow cells, leading to life-threatening shortages of blood cells.

To try to get around this problem, a doctor may treat the child with high-dose chemo (sometimes along with radiation therapy) and then use a stem cell transplant to “rescue” the bone marrow, giving the child 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 patient first gets standard doses of chemo, then local treatment of the tumor (surgery and/or radiation therapy), followed by high-dose chemo and a stem cell transplant.

**What happens in a stem cell transplant**

The first step in a stem cell transplant is to collect, or harvest, the child’s own blood-producing stem cells to use later. (These are the cells that make the different types of blood cells.) This type of transplant, where the stem cells are taken from the patient (as opposed to coming from someone else), is known as an autologous transplant.

In the past, the stem cells were often collected from the child’s bone marrow, which required a minor operation. But doctors have found that these cells can be taken from the bloodstream using a procedure similar to a blood donation. 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 child 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.

Until this happens, the child is at high risk of infection because of a low white blood cell count, as well as bleeding because of a low blood platelet count. To avoid infection, protective measures are taken, such as using special air filters in the hospital room and having visitors wear protective clothing. Blood and platelet transfusions and treatment with antibiotics may also be used to prevent or treat infections or bleeding problems.

**Practical points**

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

A stem cell transplant often requires a long hospital stay and can be very expensive (costing well over $100,000). Some insurance companies might view it as an experimental treatment and may not pay for it. Be sure to get a written approval from
your insurer before treatment if this procedure is recommended for your child. Even if the transplant is covered by your insurance, your co-pays or other costs could easily amount to many thousands of dollars. It is important to find out what your insurer will cover before the transplant to get an idea of what you might have to pay.

**Possible side effects**

The possible side effects from a stem cell transplant are generally divided into early (short-term) and late (long-term) effects.

**Early or short-term effects:** Possible early complications and side effects are basically those caused by the high-dose chemo (see the Chemotherapy section of this document) and can be severe. They can include:

- Low blood cell counts (with fatigue and increased risks of infection and bleeding)
- Nausea and vomiting
- Loss of appetite
- Mouth sores
- Diarrhea
- Hair loss

One of the most common short-term effects is an increased risk of serious infections. Antibiotics are often given to try to prevent this. Other side effects, like low red blood cell and platelet counts, might require blood product transfusions or other treatments.

**Late and long-term side effects:** Some complications and side effects can last for a long time or might not occur until years after the transplant. These can include:

- Radiation damage to the lungs
- Problems with the thyroid or other hormone-making glands
- Problems with fertility
- Cataracts (damage to the lens of the eye that can affect vision)
- Damage to bones or problems with bone growth
- Development of another cancer (including leukemia) years later

Be sure to talk to your child’s doctor before the transplant to learn about possible long-term effects your child might have.

For more on stem cell transplants, see [Stem Cell Transplant (Peripheral Blood, Bone Marrow, and Cord Blood Transplants)](Stem_Cell_Transplant_Peripheral_Blood_Bone_Marrow_and_Cord_Blood_Transplants).

- **References**
Treatment of Ewing Tumors by Stage

Treatment of a Ewing tumor is based mainly on its location and how much it has spread when first found.

Localized Ewing tumors

As mentioned earlier, even children 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 children do not get chemotherapy, these small pockets 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. It is called neoadjuvant chemotherapy because it is given before any surgery or radiation therapy. In the United States, treatment is usually a regimen known as VAdriaC/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 12 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 the tumor is not growing but surgery is not an option after the initial chemotherapy, radiation therapy (along with chemotherapy) is usually the next treatment given. In
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 is 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** is the first treatment, often using 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 at this point. Other options, such as surgery plus radiation therapy (before and/or after surgery) or radiation therapy alone to all known metastatic sites, including the lungs, 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 to try to improve the outcome for these patients. For more on this procedure, see “**High-dose chemotherapy and stem cell transplant.**”

Because these tumors can be hard to treat, clinical trials 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 recur, 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, surgery, radiation therapy, 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, as well as the use of targeted drugs and immune therapies, but it is not yet clear how useful these are. These tumors can be hard to treat, so clinical trials of newer treatments may be a good option.

- References
See all references for Ewing Family of Tumors

Social, Emotional, and Other Issues in Treating Ewing Tumors

Social and emotional issues might come up both during and after treatment. Factors such as the person’s age when diagnosed and the extent of treatment can play a role here.

Most Ewing tumors develop during the teen years, a very sensitive time in a young person’s life. A Ewing tumor and its treatment can have a profound effect on how a person looks and how they view themselves and their body. It can also affect some everyday tasks, including certain school, work, or recreational activities. These effects are often greatest during the first year of treatment, but they can last a long time in some people. It’s important that the treating center assess the family situation as soon as possible, so that any areas of concern can be addressed.

Many experts recommend that school-aged patients attend school as much as possible. This can help them maintain a sense of daily routine and keep their friends informed about what is happening.
Friends can be a great source of support, but patients and parents should know that some people have misunderstandings and fears about cancer. Some cancer centers have a school re-entry program that can help in these situations. In this program, health educators visit the school and inform students about the diagnosis, treatment, and changes that the cancer patient may go through. They also answer any questions from teachers and classmates. (For more information, see [Children Diagnosed With Cancer: Returning to School](#).)

Centers that treat many patients with Ewing tumors may have programs to introduce new patients to children or teens who have finished their treatment. This can give patients an idea of what to expect during and after treatment, which is very important. Seeing another patient with a Ewing tumor doing well can also be a source of inspiration. There are also support groups that encourage athletics and full use of the child's limbs. Many amputees or people with prosthetic limbs are able to take part in athletics and often do.

Parents and other family members can also be affected, both emotionally and in other ways. Some common family concerns during treatment include financial stresses, traveling to and staying near the cancer center, the possible loss of a job, and the need for home schooling. Social workers and other professionals at treatment centers can help families sort through these issues.

During treatment, patients and their families tend to focus on the daily aspects of getting through it and beating the cancer. But once treatment is finished, a number of emotional concerns can come up. Some of these might last a long time. They can include things like:

- Dealing with physical changes that can result from the treatment
- Worrying about the cancer returning or new health problems developing
- Feeling resentful for having had cancer or having to go through treatment when others do not
- Worrying about being treated differently or discriminated against (by friends, classmates, coworkers, employers, etc.)
- Being concerned about dating, marrying, and having a family later in life

No one chooses to have a Ewing tumor, but for many children and teens, the experience can eventually be positive, helping to establish strong self-values. Others may have a harder time recovering, adjusting to life after cancer, and moving on. It is normal to have some anxiety or other emotional reactions after treatment, but feeling overly worried, depressed, or angry can affect many parts of a young person’s growth. It can get in the way of relationships, school, work, and other aspects of life.
With support from family, friends, other survivors, mental health professionals, and others, many people who have survived cancer can thrive in spite of the challenges they’ve had to face. If needed, doctors and other members of the health care team can often recommend special support programs and services to help after cancer treatment.

Although the psychological impact of this disease in children and teens is most obvious, adults with this disease face many of the same challenges. They should also be encouraged to take advantage of the cancer center’s physical therapy, occupational therapy, and counseling services.

**References**

[See all references for Ewing Family of Tumors](#)

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