Early Detection, Diagnosis, and Staging of Ewing Tumors

Detection and Diagnosis

Catching cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that is not always the case.

- Can Ewing Tumors Be Found Early?
- Signs and Symptoms of Ewing Tumors
- Tests for Ewing Tumors

Stages and Outlook (Prognosis)

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and anticipated response to treatment.

- Ewing Tumor Stages
- Survival Rates for Ewing Tumors

Questions to Ask About Ewing Tumors

Here are some questions you can ask your cancer care team to help you better understand your cancer diagnosis and treatment options.

- Questions to Ask the Doctor About Ewing Tumors
Can Ewing Tumors Be Found Early?

Ewing tumors are uncommon, and there are no widely recommended screening tests for these cancers. (Screening is testing for cancer in people without any symptoms.) Still, Ewing tumors sometimes cause symptoms that allow them to be found early (before they have clearly spread to other parts of the body).

The most common symptom of a Ewing tumor is pain in the area of the tumor. Sometimes the tumor shows up as a lump or swelling on an arm or leg, or on the chest. Sometimes the lump feels warmer than the rest of the body, and sometimes the child has other symptoms like a fever or not feeling well.

Of course, children and teens often get sore or have lumps and bumps from normal activities. But pains or lumps that don’t go away (or that get worse) should be checked by a doctor. The same is true if a lump feels warm and/or the child has a fever. These symptoms are more likely to have other causes, such as an infection, but they need to be checked by a doctor so that the cause can be found and treated, if needed.

Hyperlinks


References


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Signs and Symptoms of Ewing Tumors

Ewing tumors are often found because of the symptoms they cause.

**Pain**

Most children and teens with Ewing tumors will have pain in the area of the tumor. Ewing tumors occur most often in the pelvis (hip bones), the chest wall (such as the ribs or shoulder blades), and the legs (mainly in the middle of the long bones), but they can also start in other parts of the body.

Bone pain can be caused by the tumor spreading under the outer covering of the bone (periosteum), or the pain can be from a break (fracture) in a bone that has been weakened by the tumor.

**Lump or swelling**

Over time, most Ewing bone tumors and almost all non-bone (soft tissue) Ewing tumors cause a lump or swelling, which is more likely to be noticed in tumors in the arms or legs. The lump is often soft and feels warm. Tumors in the chest wall or pelvis might not be noticed until they have grown quite large.

**Other symptoms**

Ewing tumors can also cause other symptoms, some of which are more common in tumors that have spread:

- Fever
- Feeling tired
- Weight loss

Rarely, tumors near the spine can cause back pain, as well as weakness, numbness, or paralysis in the arms or legs. Tumors that have spread to the lungs can cause shortness of breath.

Many of the signs and symptoms of Ewing tumors are more likely to be caused by something else. Still, if your child has any of these symptoms, see a doctor so that the cause can be found and treated, if needed.
Because many of these signs and symptoms can be confused with normal bumps and bruises or bone infections, Ewing tumors might not be recognized right away. For example, the doctor might try giving antibiotics first if an infection is suspected. The correct diagnosis might not be made until the signs and symptoms don’t go away (or get worse) and the bone is then x-rayed.

References


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Tests for Ewing Tumors

Ewing tumors are usually found because of signs or symptoms\(^1\) a person is having. If the doctor suspects a tumor, exams and tests will be needed to find out for sure.

Medical history and physical exam

If you or your child has signs or symptoms that could be from a tumor, the doctor will want to get a complete medical history to find out more about the symptoms and how long they have been present.

The doctor will also do a complete physical exam, paying special attention to any areas causing pain or swelling.

If a doctor suspects a bone tumor (or another type of tumor), more tests will be done to
find out. These might include imaging tests, biopsies, and/or lab tests.

**Imaging tests**

Imaging tests (such as x-rays, MRI scans, CT scans, bone scans, and PET scans) create pictures of the inside of the body. Imaging tests might be done for many reasons, including:

- To help find out if a suspicious area might be cancer
- To determine the extent of a tumor or learn how far a cancer may have spread
- To help determine if treatment is working

Patients who have or might have a Ewing tumor will have one or more of these tests.

**X-rays**

If a bone lump doesn’t go away or the doctor suspects a bone tumor for some other reason, an x-ray of the area will probably be the first test done. A radiologist (doctor who specializes in reading imaging tests) can usually spot a bone tumor on an x-ray and can often tell if it is likely to be a Ewing tumor. But other imaging tests might be needed as well.

Even if an x-ray strongly suggests a Ewing bone tumor, a biopsy (described below) is still needed to confirm that it is cancer rather than some other problem, such as an infection.

**Magnetic resonance imaging (MRI) scan**

MRI scans create detailed images using radio waves and strong magnets instead of x-rays, so there is no radiation involved. A contrast material called gadolinium may be injected into a vein before the scan to help see details better.

An MRI scan is often done to get a better look an abnormal area seen on an x-ray. MRI scans usually can show if it is likely to be a tumor, an infection, or some type of bone damage from other causes. MRIs can also help determine the extent of a tumor, as they show the detail inside bones as well as the muscle, fat, and connective tissue around the tumor. Knowing the extent of the tumor is very important when planning surgery or radiation therapy.

MRI scans might also be done to see if the cancer has spread to other areas, such as
the spine or pelvis (hip area). MRI scans can also be used during and after treatment to see how well the tumor is responding.

**Computed tomography (CT) scan**

A CT scan\(^4\) combines many x-ray pictures to make detailed cross-sectional images of parts of the body, including soft tissues such as muscles. A contrast material may be injected into a vein before the scan to help see details better.

CT scans of the chest are often used to see if a Ewing tumor has spread to the lungs. MRI scans are usually a bit better at defining the extent of the main tumor itself, but a CT scan of the tumor may be done as well.

**Bone scan**

For a bone scan\(^5\), a small amount of low-level radioactive material is injected into the blood and travels to the bones. A special camera can detect the radioactivity and creates a picture of the skeleton.

Areas of active bone changes appear as “hot spots” on the skeleton because they attract the radioactivity. These areas may suggest the presence of cancer, but other bone diseases can also cause the same pattern. To be sure, other tests such as plain x-rays or MRI scans, or even a bone biopsy, might be needed.

A bone scan can help show if a cancer has spread to bones in other parts of the body, and might be part of the workup for a Ewing tumor. This test is useful because it can show the entire skeleton at once. (A positron emission tomography [PET] scan can often provide similar information, so a bone scan might not be needed if a PET scan is done.)

**Positron emission tomography (PET) scan**

For a PET scan\(^6\), a form of radioactive sugar (known as FDG) is injected into the blood. Because cancer cells are growing quickly, they absorb large amounts of the sugar. A special camera can then create a picture of areas of radioactivity in the body. The picture is not detailed like a CT or MRI scan, but it provides helpful information about the whole body.

PET scans can be very helpful in showing the spread of Ewing tumors and in finding out whether abnormal areas seen on other imaging tests (such as a bone scan or CT scan) are tumors. PET scans can also be repeated during treatment to see how well it is
working.

Some newer machines can do a PET and CT scan at the same time (PET/CT scan). This lets the doctor compare areas of higher radioactivity on the PET scan with the more detailed appearance of that area on the CT scan.

**Biopsy of the tumor**

The results of imaging tests might strongly suggest a Ewing tumor, but a biopsy (removing some of the tumor for viewing under a microscope and other lab testing) is the only way to be certain.

If the tumor is in a bone, it is very important that an orthopedic surgeon experienced in treating bone tumors does the biopsy. Whenever possible, the biopsy and the surgery to treat the cancer should be planned together, and the same surgeon should do both. Proper planning of the biopsy can help prevent later complications and might reduce the amount of surgery needed later on.

There are a few ways to get a sample of the tumor to diagnose Ewing tumors.

**Incisional biopsy**

For most suspected Ewing tumors, an incisional biopsy (taking out only a piece of the tumor) is done. This can be done in a couple of ways:

- **Surgical (open) biopsy**: The surgeon cuts away a piece of the tumor through an opening on the skin.
- **Needle (closed) biopsy**: The surgeon puts a large, hollow needle through the skin and into the tumor to remove a piece of it.

Incisional biopsies are often done while the patient is under general anesthesia (in a deep sleep), but in older teens and adults they are sometimes done with just sedation and a local anesthetic (numbing medicine).

**Excisional biopsy**

In very rare cases, if the tumor is small enough and in a good location, the surgeon can completely remove it while the patient is under general anesthesia (asleep). This is called an excisional biopsy.
If general anesthesia is going to be used for the biopsy, the surgeon may also plan other procedures while the patient is asleep to avoid having to do them as separate operations later on. For example, if the tumor is thought to have spread to the chest or elsewhere, the surgeon may take biopsy samples of these suspected tumors. The doctor might also do a bone marrow biopsy (see below) at this time to see if the cancer has spread to the bone marrow.

During the biopsy (while the patient is still asleep), the biopsy samples can be checked quickly under a microscope for cancer. If it looks like a Ewing tumor, the patient will very likely need chemotherapy as part of treatment, so the surgeon may place a small flexible tube, known as a central venous catheter, into a large vein in the chest area during the same operation. The catheter end lies just under or outside on the skin. It can stay in place for several months during treatment. The catheter gives doctors and nurses easier access to the vein, so not as many needle sticks are needed to give chemotherapy or do blood draws later.

**Bone marrow aspiration and biopsy**

These tests are used to see if the cancer cells have spread into the bone marrow, the soft inner parts of certain bones. The tests might be done once a Ewing tumor has been diagnosed because it is important to know if the tumor has spread to the bone marrow.

Bone marrow aspiration and biopsy are usually done at the same time. In most cases the marrow samples are taken from the back of both of the pelvic (hip) bones. These tests may be done during the surgery to biopsy or treat the main tumor (while the patient is still under anesthesia), or they may be done as a separate procedure.

If the bone marrow aspiration is being done as a separate procedure, the patient lies on a table (on his or her side or belly). The area over the hip is cleaned, and the skin and the surface of the bone are numbed with a local anesthetic, which may cause a brief stinging or burning sensation. Children may also be given other medicines to make them sleepy, or they might even be asleep during the procedure. A thin, hollow needle is then inserted into the bone, and a syringe is used to suck out a small amount of liquid bone marrow.

A bone marrow biopsy is usually done just after the aspiration. A small piece of bone and marrow is removed with a slightly larger needle that is pushed down into the bone. Once the biopsy is done, pressure is applied to the site to help stop any bleeding.

Samples from the bone marrow are sent to a pathology lab, where they are looked at and tested for cancer cells.
Testing biopsy samples

A doctor called a **pathologist** looks at all biopsy specimens under a microscope to see if they contain cancer cells. If cancer is found, the specific type of cancer can often be determined as well. But because cells from Ewing tumors share many of the same features as cells from other types of cancer, more lab tests are often needed.

**Immunohistochemistry**

For this test, a portion of the biopsy sample is treated with special proteins (antibodies) that attach to substances found on Ewing tumor cells but not on other cancers. Chemicals (stains) are then added so that cells containing these substances change color and can be seen under a microscope. This lets the pathologist know that the cells are from a Ewing tumor.

**Chromosome tests**

Normal human cells have 23 pairs of chromosomes (strands of DNA), each of which is a certain size and looks a certain way under the microscope. Ewing tumor cells almost always have chromosome translocations, where 2 chromosomes swap pieces of their DNA. In most cases, the cells have translocations between chromosomes 22 and 11. Less often, the translocation is between other chromosomes. Finding these changes can help doctors tell Ewing tumors from other types of cancer. Other types of chromosome changes can also be found in some Ewing tumors.

**Cytogenetics:** In this lab test, the cells are looked at under a microscope to see if the chromosomes have any abnormalities. A drawback of this test is that it usually takes about 2 to 3 weeks because the cells must grow in lab dishes for a couple of weeks before their chromosomes are ready to be viewed under the microscope.

**Fluorescent in situ hybridization (FISH):** This test looks more closely at tumor cell DNA using special fluorescent dyes that only attach to specific genes or parts of chromosomes. FISH can find most chromosome changes (such as translocations) that can be seen in standard cytogenetic tests, as well as some gene changes too small to be seen with cytogenetic testing. FISH is very accurate and can usually provide results within a couple of days.

**Polymerase chain reaction (PCR):** PCR is a very sensitive test that is often able to detect very small numbers of cells with translocations, which wouldn’t be detected by cytogenetics.
PCR is also useful in looking for cancer remaining or coming back after treatment. For example, if PCR testing of a bone marrow sample after treatment finds cells with a typical Ewing tumor translocation, it’s likely that the cancer hasn’t been cured, and that more treatment is needed.

**Blood tests**

No blood test can be used to diagnose Ewing tumors. But certain blood tests may be helpful once a diagnosis has been made.

A **complete blood count (CBC)** measures the levels of white blood cells, red blood cells, and platelets in the blood. An abnormal CBC result at the time of diagnosis might suggest the cancer has spread to the bone marrow, where these blood cells are made.

A blood test for levels of an enzyme called **lactate dehydrogenase (LDH)** is typically done at diagnosis. A high LDH level is often a sign that there is more cancer in the body.

Standard blood tests are done often to check a patient’s general health both before treatment (especially before surgery) and during treatment (such as chemotherapy) to look for possible problems or side effects. These tests often include a CBC to monitor bone marrow function and **blood chemistry tests** to measure how well the liver and kidneys are working.

**Hyperlinks**

Ewing Tumor Stages

Once a Ewing tumor has been diagnosed, tests are done to determine the stage (extent of spread) of the cancer. The stage of a Ewing tumor describes how much cancer is in the body. It helps determine how serious the cancer is and how best to treat it. Doctors also use a cancer’s stage when talking about survival statistics.

The stage is based on results of imaging tests and biopsies of the main tumor and other tissues, which are described in Tests for Ewing Tumors.
A **staging system** is a standard way for the cancer care team to sum up the extent of the cancer. The formal (and more detailed) staging system for Ewing tumors is the American Joint Committee on Cancer (AJCC) system for bone cancer. It is described below to help you understand it, in case your doctor refers to it. But for treatment purposes, doctors often use a simpler system, dividing Ewing tumors into 2 groups: localized or metastatic.

Staging can be confusing. If you have any questions about the stage of the cancer, ask someone on the health care team to explain it to you in a way you understand.

**Localized vs. metastatic stages**

When determining how best to treat a Ewing tumor, doctors typically classify them as either localized or metastatic.

**Localized Ewing tumors**

Doctors call a Ewing tumor "localized" if they believe it's only in the area where it started or in nearby tissues such as muscle or tendons. A Ewing tumor is considered localized only after all of the imaging tests (x-rays, CT or MRI scans, and PET or bone scans) and the bone marrow biopsy and aspirate (if done) do not find it has spread to distant parts of the body.

Even when imaging tests do not show that the cancer has spread to distant areas, most patients are likely to have **micrometastases** (very small areas of cancer spread that can’t be detected with tests). This is why chemotherapy, which can reach all parts of the body, is an important part of treatment for all Ewing tumors.

**Metastatic Ewing tumors**

A metastatic Ewing tumor has clearly spread from where it started to distant parts of the body. Most of the time, it spreads to the lungs or to other bones or the bone marrow. Less commonly, it spreads to the liver or lymph nodes.

About 1 in 5 patients will have obvious spread that is found by imaging tests. But as mentioned above, many other patients are likely to have small amounts of cancer spread to other parts of the body that can’t be seen on imaging tests.

**AJCC staging system for bone cancer**
The AJCC uses one system to describe all bone cancers, including Ewing tumors that start in bone.

*Extraosseous Ewing (EOE) tumors (Ewing tumors that don’t start in bones) are staged differently. They are staged like soft tissue sarcomas. Information about soft tissue sarcoma staging can be found in Sarcoma - Adult Soft Tissue Cancer*.  

The AJCC staging system for bone cancers is based on 4 key pieces of information:

- **T** describes the size of the main (primary) **tumor** and whether it appears in different areas of the bone.
- **N** describes the extent of spread to nearby (regional) lymph **nodes** (small bean-sized collections of immune system cells). Bone tumors rarely spread to the lymph nodes.
- **M** indicates whether the cancer has **metastasized** (spread) to other organs of the body. (The most common sites of spread are to the lungs or other bones.)
- **G** stands for the **grade** of the tumor, which describes how the cells from biopsy samples look. Low-grade tumor cells look more like normal cells and are less likely to grow and spread quickly, while high-grade tumor cells look more abnormal. (All Ewing tumors are considered high-grade tumors.)

Numbers or letters after T, N, M, and G provide more details about each of these factors.

**T categories of bone cancer**

- T0: There is no evidence of a main (primary) tumor.  
- T1: The tumor is no more than 8 cm (around 3 inches) across.  
- T2: The tumor is larger than 8 cm across.  
- T3: The tumor is in more than one site in the same bone.

*The T categories are slightly different if the main tumor is in the pelvis (hip bone) or spine.

**N categories of bone cancer**

- N0: There is no spread to nearby lymph nodes.  
- N1: The cancer has spread to nearby lymph nodes.
M categories of bone cancer

- M0: There is no spread (metastasis) to distant organs.
- M1a: The cancer has spread only to the lungs.
- M1b: The cancer has spread to other distant parts of the body.

Grades of bone cancer

- GX: Grade can’t be assessed
- G1: Low grade
- G2-G3: High grade

(All Ewing tumors are considered G3.)

Stage grouping

Once the T, N, and M categories and the grade of the bone cancer have been determined, the information is combined and expressed as an overall stage. The process of assigning a stage number is called stage grouping. The stages are described in Roman numerals from I to IV (1-4), and are sometimes divided further.

Stage IA*

T1, N0, M0, G1 (or GX): The tumor is no more than 8 cm across (T1) and is low grade (or the grade can’t be assessed). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).

Stage IB*

T2 or T3, N0, M0, G1 (or GX): The tumor is either larger than 8 cm across (T2) or it is in more than one place in the same bone (T3). It is low grade (or the grade can’t be assessed). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).

Stage IIA

T1, N0, M0, G2 to G3: The tumor is no more than 8 cm across (T1) and is high grade (G2 or G3). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).
Stage IIB

**T2, N0, M0, G2 to G3:** The tumor is larger than 8 cm across (T2) and is high grade (G2 or G3). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).

Stage III

**T3, N0, M0, G2 to G3:** The tumor is in more than one place in the same bone (T3). It is high grade (G2 or G3). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).

Stage IVA

**Any T, N0, M1a, any G:** The tumor has spread only to the lungs (M1a). It has not spread to the lymph nodes (N0) or to other parts of the body. (It can be any size or grade.)

Stage IVB (if either of these applies)

**Any T, N1, any M, any G:** The tumor has spread to nearby lymph nodes (N1). It can be any size or grade, and may or may not have spread to other parts of the body.

**Any T, any N, M1b, any G:** The tumor has spread to distant parts of the body other than the lungs (M1b). It can be any size or grade.

*All Ewing tumors are classified as G3 (high grade), so they are never stage I bone cancers.*

Hyperlinks


References
Survival Rates for Ewing Tumors

Survival rates are often used by doctors as a standard way of discussing a person’s prognosis (outlook). These numbers tell you what portion of people with the same type and stage of cancer are still alive a certain amount of time (usually 5 years) after they were diagnosed. They can’t tell you exactly what will happen with any person, but they may help give you a better understanding about how likely it is that treatment will be successful. Some people find survival rates helpful, but some people might not.

What is a 5-year survival rate?

Statistics on the outlook for a certain type and stage of cancer are often given as 5-year survival rates. The 5-year survival rate is the percentage of people who live at least 5 years after being diagnosed with cancer. For example, a 5-year survival rate of 70% means that an estimated 70 out of 100 people who have that cancer are still alive 5 years after being diagnosed. Of course, many people live much longer than 5 years (and many are cured).

But remember, the 5-year survival rates are estimates – each person’s outlook can vary
based on a number of factors specific to them.

Cancer survival rates don’t tell the whole story

Survival rates are often based on previous outcomes of large numbers of people who had the disease, but they can’t predict what will happen in any person’s case. There are some important limitations to remember:

- The 5-year survival rates below are among the most current available. But to get these numbers, doctors have to look at people who were treated at least 5 years ago. As treatments are improving over time, people who are now being diagnosed with Ewing tumors may have a better outlook than these statistics show.
- These statistics are based on the extent of the cancer when it was first diagnosed. They do not apply to cancers that later come back or spread, for example.
- The outlook for people with Ewing tumors varies by the stage (extent) of the cancer – in general, the survival rates are higher for people with earlier stage cancers. But many other factors can also affect a person’s outlook (see below).

Your cancer care team can tell you how the numbers below may apply, as they are familiar with your (child’s) particular situation.

Survival rates for Ewing tumors, by stage

Localized tumors

Ewing tumors are considered to be localized if there is no obvious cancer spread to other parts of the body (even if there might be small areas of spread that can't be found on tests). With current treatment, the overall 5-year survival rate for patients with Ewing tumors that are still localized when they are first found is around 70%.

Metastatic tumors

If the cancer has clearly already spread when it is diagnosed, the 5-year survival rate is about 30%. The survival rate is slightly better if the cancer has only spread to the lungs as opposed to having reached other organs.

Other factors affecting prognosis
Factors other than the stage of the cancer can also affect survival rates. Factors that have been linked with a better prognosis include:

- Smaller tumor size
- Main tumor is on an arm or leg (as opposed to chest wall or pelvis)
- Normal blood LDH$^2$ level
- Good tumor response to chemotherapy$^3$
- Age younger than 10 years

Even when taking these other factors into account, survival rates are at best rough estimates. Your cancer care team is your best source of information on this topic.

Hyperlinks


References


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Questions to Ask the Doctor About Ewing Tumors

It’s important to have honest, open discussions with your cancer care team. Ask any question, no matter how minor it might seem. For instance, consider these questions:

**Before getting a bone biopsy**

- How much experience do you have doing this type of biopsy?
- Are you part of a team that treats bone cancers?
- What will happen during the biopsy?
- How long will it take to get the results from the biopsy?

**If a Ewing tumor has been diagnosed**

- What kind of Ewing tumor do I (does my child) have?
- Where exactly is the tumor?
- Has the cancer spread beyond where it started?
- What is the stage\(^1\) of the cancer and what does that mean?
- Do we need any other tests\(^2\) before we can decide on treatment?
- How much experience do you have treating this type of cancer?
- Who else will be on the treatment team, and what do they do?

**When deciding on a treatment plan**

- What are our treatment options\(^3\)?
- What do you recommend and why?
- Are there any clinical trials\(^4\) we should consider? How can we find out more about them?
- What’s the goal of treatment?
- Should we get a second opinion\(^5\)? How do we do that? Can you recommend someone?
- How soon do we need to start treatment?
- What should I (we) do to be ready for treatment?
- How long will treatment last? What will it be like? Where will it be done?
How will treatment affect our daily lives?
What are the risks and side effects of the suggested treatments?
Which side effects start shortly after treatment and which ones might develop later on?
Will treatment affect my child’s ability to grow and develop?
Will treatment affect my child’s future ability to have children?

During and after treatment

Once treatment begins, you’ll need to know what to expect and what to look for. Not all of these questions may apply, but getting answers to the ones that do may be helpful.

- How will we know if the treatment is working?
- Is there anything we can do to help manage side effects?
- What symptoms or side effects should we tell you about right away?
- How can we reach you or someone on your team on nights, weekends, or holidays?
- Who can we talk to if we have questions about costs, insurance coverage, or social support?
- What are the chances of the cancer coming back after treatment? What might our options be if this happens?
- What type of follow up and rehab will be needed after treatment?
- Are there nearby support groups or other families who have been through this that we could talk to?

Along with these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery times so that you can plan your work or school schedules.

Keep in mind that doctors aren’t the only ones who can give you information. Other health care professionals, such as nurses and social workers, can answer some of your questions. To find more about speaking with your health care team, see The Doctor-Patient Relationship.

Hyperlinks


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