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Early Detection, Diagnosis, and Staging of Ewing Tumors

Learn about the signs and symptoms of Ewing tumors. Find out how Ewing tumors are tested for, diagnosed, and staged.

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 s99)

Can Ewing Tumors Be Found Early?

Ewing tumors (Ewing sarcomas) 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 or teen has other symptoms like a fever or not feeling well.

Of course, children and teens might get sore areas or 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.

References

Anderson ME, Dubois SG, Gebhart MC. Chapter 89: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepp/ gs (h 12 Tf and bu0 0 Td 0 /.dl 72 lgs

Signs and Symptoms of Ewing Tumors

- Pain
- Lump or swelling

Rarely, tumors near the spine can affect nearby nerves, which can lead to **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, a person with any of these symptoms, especially if they don't go away or get worse, should see a doctor so 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 with 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 painful area is then x-rayed.

References

Anderson ME, Dubois SG, Gebhart MC. Chapter 89: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 6th ed. Philadelphia, Pa: Elsevier; 2020.

DeLaney TF, Hornicek FJ. Clinical presentation, staging, and prognostic factors of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/clinical-presentation-staging-and-prognostic-factors-of-theewing-sarcoma-family-of-tumors on October 30, 2020.

National Cancer Institute. Ewing Sarcoma Treatment (PDQ). 2020. Accessed at https://www.cancer.gov/types/bone/patient/ewing-treatment-pdq October 30, 2020.

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Tests for Ewing Tumors

- Medical history and physical exam
- Imaging tests
- Biopsy of the tumor
- Bone marrow aspiration and biopsy

- Testing biopsy samples
- Blood tests

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

If a Ewing tumor is found, other tests will then be needed to learn more about it.

Medical history and physical exam

If a person has signs or symptoms that could be from a bone or soft tissue 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 **physical exam**, paying special attention to any areas causing pain or swelling.

If the doctor suspects a bone tumor (or another type of tumor), more tests will be done. 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

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

X-rays

If a lump in or near a bone doesn't go away or if the doctor suspects a bone tumor for some other reason, an x-ray of the area will probably be the first test done. Doctors 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 image strongly suggests a Ewing tumor, a biopsy (described below) will still be needed to confirm that it is cancer rather than some other problem, such as an infection.

Magnetic resonance imaging (MRI) scan

MRIs² create detailed images using radio waves and strong magnets instead of x-rays, so there is no radiation involved. A contrast material called **gadolinium** is often injected into a vein before the scan to help see details better.

A bone scan can help show if a cancer has spread to bones in other parts of the body, so it 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 scan⁵, a form of radioactive sugar (known as **FDG**/GSt55iiiiB0jbjuroo bhe boitron 0.2 0g

- 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

the marrow samples are taken from the back of the pelvic (hip) bones. These tests may be done during surgery to biopsy or treat the main tumor (while the patient is still under

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 with 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 looked at with a 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 that remains or comes 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 at least some cancer remains, and that more treatment is needed.

Genomics testing: Some newer lab tests can look at all of the genes inside cancer cells at the same time. These tests, also known as **next generation sequencing**, can sometimes help guide treatment of the cancer.

Blood tests

No blood test can be used to diagnose Ewing tumors. But certain <u>blood tests</u>⁹ 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 (that is, low blood cell counts) 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 <u>surgery</u>¹⁰) and during treatment (such as <u>chemotherapy</u>¹¹) to look for possible problems or side effects. These tests often include a CBC to check blood cell levels and blood chemistry tests to measure how well the liver and kidneys are working.

Hyperlinks

- 1. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/x-rays-and-other-radiographic-tests.html</u>
- 2. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/mri-for-cancer.html
- 3. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ct-scan-for-cancer.html</u>
- 4. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/nuclear-medicine-scans-for-cancer.html</u>
- 5. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/nuclear-medicine-scans-for-cancer.html</u>
- 6. www.cancer.org/cancer/types/ewing-tumor/treating/surgery.html
- 7. www.cancer.org/cancer/types/ewing-tumor/treating/chemotherapy.html
- 8. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/tubes-lines-ports-catheters.html</u>
- 9. <u>www.cancer.org/cancer/diagnosis-staging/tests/understanding-your-lab-test-results.html</u>
- 10. www.cancer.org/cancer/types/ewing-tumor/treating/surgery.html
- 11. www.cancer.org/cancer/types/ewing-tumor/treating/chemotherapy.html

References

Anderson ME, Dubois SG, Gebhart MC. Chapter 89: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 6th ed. Philadelphia, Pa: Elsevier; 2020.

UpToDate. 2020. Accessed at https://www.uptodate.com/contents/bone-tumors-diagnosis-and-biopsy-techniques on January 27, 2021.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2021. Accessed at www.nccn.org/professionals/physician_gls/pdf/bone.pdf on January 27, 2021.

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Ewing Tumor Stages

- Localized vs. metastatic Ewing tumors
- AJCC staging system for bone cancers
- AJCC staging system for soft tissue sarcomas

Once a Ewing tumor (Ewing sarcoma) has been diagnosed, tests are done to determine the **stage** (extent) 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 treat¹ 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 any other body 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. Different types of cancers have different staging systems.

The formal (and more detailed) staging systems for Ewing tumors are the American Joint Committee on Cancer (AJCC) systems for bone cancers and for soft tissue sarcomas. They are described briefly below to help you understand them, in case your doctor refers to one of them.

But for treatment purposes, doctors often use a simpler system, describing Ewing tumors as either localized or metastatic.

Localized vs. metastatic Ewing tumors

When determining how best to treat Ewing tumors, doctors typically classify them as

either localized or metastatic.

Localized Ewing tumors

Doctors call a Ewing tumor localized if it can only be detected in the area where it started or in nearby tissues such as muscle or tendons. A Ewing tumor is considered localized only after all tests have been done (including imaging tests such as x-rays, CT or MRI scans, and PET or bone scans, and possibly a bone marrow biopsy), and they don't show the cancer has spread to distant parts of the body.

Even when imaging tests don't 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 <u>chemotherapy</u>², 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 cancers

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

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 beansized 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 [G3] tumors.)

Once the T, N, M, and G categories 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 to 4), and are sometimes divided further. For more detailed information on the AJCC TNM staging system for bone cancers, see <u>Bone Cancer Stages</u>³.

AJCC staging system for soft tissue sarcomas

Extraosseous Ewing (EOE) tumors (Ewing tumors that don't start in bones) are staged like soft tissue sarcomas. The AJCC staging system for soft tissue sarcomas is based on 4 key pieces of information:

- T describes the size of the main (primary) tumor.
- N describes the extent of spread to nearby (regional) lymph **nodes** (small beansized collections of immune system cells).
- M indicates whether the cancer has metastasized (spread) to other organs of the body.
- **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 [G3] tumors.)

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

Once the T, N, M, and G categories 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 to 4), and are sometimes divided further. For more information about soft tissue sarcoma staging, see Soft Tissue Sarcoma Stages⁴.

Ewing tumor stages can be confusing, so be sure to ask someone on the health care team if you have any questions about the stage of the cancer.

Hyperlinks

- 1. www.cancer.org/cancer/types/ewing-tumor/treating.html
- 2. www.cancer.org/cancer/types/ewing-tumor/treating/chemotherapy.html
- 3. <u>www.cancer.org/cancer/types/bone-cancer/detection-diagnosis-staging/staging.html</u>
- 4. www.cancer.org/cancer/types/soft-tissue-sarcoma/detection-diagnosis-staging/staging.html

References

American Joint Committee on Cancer. Bone. In: *AJCC Cancer Staging Manual*. 8th ed. New York, NY: Springer; 2017: 471-486.

American Joint Committee on Cancer. Soft Tissue Sarcoma of the Trunk and Extremities. In: *AJCC Cancer Staging Manual*. 8th ed. New York, NY: Springer; 2017:507.

Anderson ME, Dubois SG, Gebhart MC. Chapter 89: Sarcomas of bone. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 6th ed. Philadelphia, Pa: Elsevier; 2020.

DeLaney TF, Hornicek FJ. Clinical presentation, staging, and prognostic factors of the Ewing sarcoma family of tumors. UpToDate. Accessed at www.uptodate.com/contents/clinical-presentation-staging-and-prognostic-factors-of-the-ewing-sarcoma-family-of-tumors on January 28, 2021.

National Comprehensive Cancer Network (NCCN). Practice Guidelines in Oncology: Bone Cancer. Version 1.2021. Accessed at www.nccn.org/professionals/physician_gls/pdf/bone.pdf on January 28, 2021.

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Survival Rates for Ewing Tumors

- What is a 5-year relative survival rate?
- Where do these numbers come from?
- 5-year relative survival rates for Ewing tumors
- Understanding the numbers
- Other factors that can affect prognosis (outlook)

Survival rates can give you an idea of what percentage 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 hol rates fo 1 87.6 656 Tm /F3 4.5 Tf 0 0 0 (and 0.6ci. 1 0 0 1 72 5

structures, or it has reached nearby lymph nodes.

• **Distant:** The cancer has spread to distant parts of the body, such as to the lungs or to bones in other parts of the body.

5-year relative survival rates for Ewing tumors

These numbers are based on people diagnosed with Ewing tumors between 2012 and 2018.

SEER* stage	5-year relative survival rate
Localized	82%
Regional	71%
Distant	39%
All SEER stages combined	63%

^{*}SEER = Surveillance, Epidemiology, and End Results

Understanding the numbers

- These numbers apply only to the stage of the cancer when it is first diagnosed. They do not apply later on if the cancer grows, spreads, or comes back after treatment.
- These numbers don't take everything into account. Survival rates are grouped based on how far the cancer has spread. But other factors, such as those listed below, can also affect a person's outlook.
- People now being diagnosed with Ewing tumors may have a better outlook than these numbers show. Treatments improve over time, and these numbers are based on people who were diagnosed and treated at least 5 years earlier.

Other factors that can affect prognosis (outlook)

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

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Questions to Ask About Ewing Tumors

- Before getting a bone biopsy
- If a Ewing tumor (Ewing sarcoma) has been diagnosed
- When deciding on a treatment plan
- During and after treatment

It's important to have honest, open discussions with your health 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 (Ewing sarcoma) has been diagnosed

- What kind of Ewing tumor is it?
- Where exactly is the tumor?
- Has the cancer spread beyond the place where it started?
- What is the stage of the cancer, and what does that mean?
- Do we need any other tests before we can decide on treatment?
- Will we need to see any other types of doctors?
- 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 options1?

- What do you recommend and why?
- Are there any <u>clinical trials</u>² we should consider? How can we find out more about them?
- What's the goal of treatment?
- Should we get a <u>second opinion</u>³? How do we do that? Can you recommend a doctor or cancer center?
- How soon do we need to start treatment?
- What should I (we) do to be <u>ready for treatment</u>⁴?
- 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?
- Are there <u>fertility issues</u>⁵we need to consider?

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 <u>follow up</u>⁶ and rehab will be needed after treatment?
- Do you know of any local or online support groups where we can talk to other families who have been through this?

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.

Other health care professionals, such as nurses and social workers, also can answer some of your questions. To find more about speaking with your health care team, see The Doctor-Patient Relationship⁷.

Hyperlinks

- 1. www.cancer.org/cancer/types/ewing-tumor/treating.html
- 2. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-trials.html</u>
- 3. <u>www.cancer.org/cancer/managing-cancer/finding-care/seeking-a-second-opinion.html</u>
- 4. www.cancer.org/cancer/managing-cancer/making-treatment-decisions.html
- 5. <u>www.cancer.org/cancer/managing-cancer/side-effects/fertility-and-sexual-side-effects.html</u>
- 6. www.cancer.org/cancer/types/ewing-tumor/follow-up.html
- 7. <u>www.cancer.org/cancer/managing-cancer/finding-care/the-doctor-patient-relationship.html</u>

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