About Ewing Tumors

Overview and Types

If you or your child have just been diagnosed with a Ewing tumor or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Is the Ewing Family of Tumors?

Research and Statistics

See the latest estimates for new cases of Ewing tumors in the US and what research is currently being done.

- Key Statistics for Ewing Tumors

What Is the Ewing Family of Tumors?

For information about the differences between childhood cancers and adult cancers, see Cancer in Children[2].

The main types of Ewing tumors are:

- **Ewing sarcoma of bone:** Ewing sarcoma that starts in a bone is the most common tumor in this family. This type of tumor was first described by Dr. James Ewing in 1921, who found it was different from the more common bone tumor, osteosarcoma[3]. Seen with a microscope, its cells looked different from osteosarcoma cells. It was also more likely to respond to radiation therapy.
- **Extraosseous Ewing tumor (EOE):** Extraosseous Ewing tumors start in soft tissues around bones, but they look and act very much like Ewing sarcomas in bones. They are also known as **extraskeletal** Ewing sarcomas.
- **Peripheral primitive neuroectodermal tumor (PPNET):** This rare childhood cancer also starts in bone or soft tissue and shares many features with Ewing sarcoma of bone and EOE. Peripheral PNETs that start in the chest wall are known as **Askin tumors**.

Researchers have found that the cells that make up Ewing sarcoma, EOE, and PPNET are very similar. They tend to have the same DNA (gene) abnormalities and share similar proteins, which are rarely found in other types of cancers. That’s why these cancers are thought to develop from the same type of cells in the body. There are slight differences among these tumors, but they're all treated[4] the same way.

Most Ewing tumors occur in the bones. The most common sites are:

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Key Statistics for Ewing Tumors

Ewing tumors (Ewing sarcomas) are not common. About 1% of all childhood cancers are Ewing tumors. About 200 children and teens are diagnosed with Ewing tumors (sarcomas) in the United States each year.

Most Ewing tumors occur in teens, but they can also affect younger children, as well asLas_ t they car

rates.html
2. cancerstatisticscenter.cancer.org/

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What's New in Ewing Tumor Research and Treatment?

Research on Ewing tumors (Ewing sarcomas) is being done at many medical centers, university hospitals, and other institutions across the world.

Understanding and diagnosing Ewing tumors

Scientists are developing new techniques to more accurately diagnose Ewing tumors. New lab tests[1] of tumor samples are being studied to see if they can help better identify Ewing tumors and give more information on how well treatments might work against that particular tumor.

As an example, advances in technology are now allowing doctors to do **genomic testing**, which looks at all of the genes inside the cancer cells, to better understand what makes these cells different from normal cells. Some of these differences might prove to be useful targets for developing new treatments for Ewing tumors (see below).

Treatment

Researchers are looking to develop better treatments for Ewing tumors, as well as to find less toxic treatments for those that can be cured.

Radiation therapy

cancer, and doctors are now studying how to best use these tests to guide the choice of treatment. It might also lead to new drugs that target these changes in Ewing tumor cells.

Some new drugs that target specific changes in Ewing tumor cells are already being tested. For example, **TK216** is a drug that targets the main fusion protein in Ewing tumor cells that is thought to help these cells grow. Early studies of this drug in people with Ewing tumors are now under way.

Also being studied in clinical trials[7] are drugs that target the insulin-like growth factor receptor-1 (IGF-1R), a protein on some cancer cells that causes them to grow. Early studies have found that drugs like this, such as **ganitumab**, can shrink some Ewing tumors and slow down the growth of others. So far, this benefit has been temporary in most cases. These drugs may work best when combined with other drugs, which is now being tested.

Other drugs being studied for use against Ewing tumors include:

- Drugs that target certain proteins that help tumors grow and make new blood vessels, such as **cabozantinib (Cabometyx)** and **regorafenib (Stivarga)**

1. www.cancer.org/cancer/ewing-tumor/detection-diagnosis-staging/how-diagnosed.html
2. www.cancer.org/cancer/ewing-tumor/treating/radiation-therapy.html
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