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About Osteosarcoma

Get an overview of osteosarcoma and the latest key statistics in the US.

Overview and Types

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

What Is Osteosarcoma?

Research and Statistics

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

Key Statistics for Osteosarcoma

What Is Osteosarcoma?

most common).

- Osteoblastic
- Chondroblastic
- Fibroblastic
- Small cell
- Telangiectatic
- High-grade surface (juxtacortical high grade)

Other high-grade osteosarcomas include:

- Pagetoid: a tumor that develops in someone with <u>Paget disease of the bone</u>³
- Extraskeletal: a tumor that starts in a part of the body other than a bone (but still makes bone tissue)
- Post-radiation: a tumor that starts in a bone that had once been exposed to radiation

Intermediate-grade osteosarcomas

These uncommon tumors fall between high-grade and low-grade osteosarcomas. (They are usually <u>treated</u>⁴ the same way as low-grade osteosarcomas.)

Periosteal (juxtacortical intermediate grade)

Low-grade osteosarcomas

These are the slowest-growing osteosarcomas. The tumors look more like normal bone and have few dividing cells when seen with a microscope.

- Parosteal (juxtacortical low grade)
- Intramedullary or intraosseous well differentiated (low-grade central)

The grade of the tumor plays a role in determining its stage and the type of treatment used. For more on staging, see <u>Osteosarcoma Stages</u>⁵.

Other types of bone tumors

Several other types of tumors can start in the bones.

Malignant (cancerous) bone tumors

Ewing tumors (Ewing sarcomas) are the second most common bone cancer in children. They are described in <u>Ewing Family of Tumors</u>⁶.

Most other types of bone cancers are usually found in adults and are uncommon in children. These include:

- Chondrosarcoma (cancer that develops from cartilage)
- Undifferentiated pleomorphic sarcoma (UPS) of bone, previously known as malignant fibrous histiocytoma (MFH) of bone
- · Fibrosarcoma of bone
- Chordoma
- · Malignant giant cell tumor of bone

For more information on these cancers, see <u>Bone Cancer in Adults</u>⁷.

Many types of cancer that start in other organs of the body, especially cancers in adults, can spread to the bones. These are sometimes referred to as **metastatic bone cancers**, but they are not true bone cancers. For example, prostate cancer that spreads to the bones is still prostate cancer and is treated like prostate cancer. For more information, see <u>Bone Metastasis</u>⁸.

Benign (non-cancerous) bone tumors

Not all bone tumors are cancer. Benign bone tumors do not spread to other parts of the body. They are usually not life threatening, and surgery can often remove them completely. There are many types of benign bone tumors, including:

- Osteoma
- Chondroma
- Osteochondroma
- Eosinophilic granuloma of bone
- Non-ossifying fibroma
- Enchondroma
- Benign giant cell tumor of bone
- Lymphangioma

Hyperlinks

- 1. <u>www.cancer.org/cancer/understanding-cancer/what-is-cancer.html</u>
- 2. www.cancer.org/cancer/types/cancer-in-children.html
- 3. <u>www.cancer.org/cancer/types/osteosarcoma/causes-risks-prevention/risk-factors.html</u>
- 4. www.cancer.org/cancer/types/osteosarcoma/treating/by-extent.html
- 5. <u>www.cancer.org/cancer/types/osteosarcoma/detection-diagnosis-staging/staging.html</u>
- 6. www.cancer.org/cancer/types/ewing-tumor.html
- 7. www.cancer.org/cancer/types/bone-cancer.html
- 8. www.cancer.org/cancer/managing-cancer/advanced-cancer/bone-metastases.html

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Key Statistics for Osteosarcoma

Osteosarcoma is not a common cancer. Each year, about 1,000 new cases of osteosarcoma are diagnosed in the United States. About half of these are in children and teens.

Most osteosarcomas occur in children, teens, and young adults between the ages of 10 and 30. Teens are the most commonly affected age group, but people of any age can develop osteosarcoma. About 1 in 10 osteosarcomas occur in people older than 60.

About 2% of childhood cancers are osteosarcomas, but they make up a much smaller percentage of adult cancers.

The prognosis (outlook) for people with osteosarcoma depends on many factors, including where the tumor is, if the cancer has already spread (metastasized) when it's first found, and the person's age. For more on this, see <u>Survival Rates for</u> Osteosarcoma.¹

Visit the <u>American Cancer Society's Cancer Statistics Center</u>² for more key statistics.

Hyperlinks

- www.cancer.org/cancer/types/osteosarcoma/detection-diagnosis-staging/survivalrates.html
- 2. cancerstatisticscenter.cancer.org/

References

American Cancer Society. *Cancer Facts & Figures 2020*. Atlanta, Ga. American Cancer Society; 2020.

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Treatment

Many advances have been made in treating osteosarcoma in the past few decades. Still, more research is needed to learn how best to manage hard-to-treat osteosarcomas, such as those that have already spread when they are found. Many clinical trials are focusing on treating osteosarcoma using a variety of strategies.

Surgery

or slightly higher dose. This is called fractionated stereotactic radiotherapy.

Another newer approach is to use radioactive particles instead of x-rays to deliver the radiation. One example is **conformal proton beam therapy**, which uses positive parts of atoms. Unlike x-rays, which release energy both before and after they hit their target, protons cause little damage to normal 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 to do less damage to nearby normal tissues. Proton beam therapy may be helpful for hard-to-treat tumors, such as those on the spine or pelvic bones, but only a limited number of centers in the United States offer this treatment at this time.

An even newer approach uses **carbon ions**, which are heavier than protons and cause more damage to cancer cells. This therapy is still in the earliest stages of development and is only available in a small number of centers around the world.

Doctors are also studying newer forms of **radioactive drugs** to treat osteosarcoma that has spread to many bones. One example is radium-223 (Xofigo), which works slightly differently than the other radioactive drugs now being used.

Chemotherapy

Clinical trials are being done to determine the best combinations of <u>chemotherapy</u>³ (chemo) drugs, as well as the best time to give them. Newer chemo drugs are being studied as well.

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- Drugs called <u>immune checkpoint inhibitors</u>⁵ can sometimes help the body's immune system recognize and attack cancer cells. These drugs have already been shown to be helpful against many types of cancer, and some of them are now being studied for use against osteosarcoma.
- An experimental immune-stimulating drug called muramyl tripeptide (also known as MTP or mifamurtide) has been shown to help some patients when added to chemotherapy.
- Monoclonal antibodies⁶ are man-made versions of immune system proteins that attach to a specific target in the body, which can help the immune system find and destroy cancer cells. Antibodies directed against GD2 and other substances on osteosarcoma cells are now being tested in clinical trials.
- Researchers are also studying a newer form of immunotherapy known as <u>CAR-T</u> <u>cell therapy</u>⁷ for osteosarcoma that is no longer helped by other treatments.

Targeted therapy drugs

Doctors are also studying new medicines that target specific molecules on or in cancer cells. These are known as <u>targeted therapies</u>⁸.

Monoclonal antibodies, discussed above, can also be considered a type of targeted therapy. These antibodies attach to certain substances on cancer cells, which can kill them or help to stop their growth. An example is **dinutuximab** (Unituxin), an antibody that attaches to GD2, a substance that is important for cancer cell growth.

Many other targeted drugs are being studied for use against osteosarcoma, including drugs that affect a tumor's ability to make new blood vessels, such as sorafenib (Nexavar), pazopanib (Votrient), lenvatinib (Lenvima), and cabozantinib (Cabometyx).

Drugs that affect the bones

Drugs that target bone cells called osteoclastsmay also be useful against osteosarcoma:

- Bisphosphonates are a group of drugs that are already used to treat osteoporosis (bone thinning) and certain cancers that have spread to the bones. Some of these drugs, such as pamidronate and zoledronic acid, are now being studied for use in osteosarcoma as well.
- **Denosumab** is a monoclonal antibody that targets the RANKL protein, which normally helps bones grow. It is now being studied for use against osteosarcoma.

Childhood Cancer Research Highlights 9

The American Cancer Society is committed to finding new answers to help every child and family affected by cancer--see some of our latest research.

Hyperlinks

- 1. www.cancer.org/cancer/types/neuroblastoma.html
- 2. www.cancer.org/cancer/types/osteosarcoma/treating/radiation-therapy.html
- 3. www.cancer.org/cancer/types/osteosarcoma/treating/chemotherapy.html
- 4. www.cancer.org/cancer/managing-cancer/advanced-cancer/lung-metastases.html
- 5. <u>www.cancer.org/cancer/managing-cancer/treatment-types/immunotherapy/immune-checkpoint-inhibitors.html</u>
- 6. <u>www.cancer.org/cancer/managing-cancer/treatment-types/immunotherapy/monoclonal-antibodies.html</u>
- 7. <u>www.cancer.org/cancer/managing-cancer/treatment-types/immunotherapy/car-t-cell1.html</u>
- 8. <u>www.cancer.org/cancer/managing-cancer/treatment-types/targeted-therapy.html</u> www.cancer.org/research/acs-research-highlights/childhood-cancer-research-

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