

OSTEOSARCOMA

**A PUBLICATION OF
THE BONE AND CANCER FOUNDATION**



DEFINITION

1. What is osteosarcoma?

Osteosarcoma is a cancerous tumor that occurs in the bone.

Most osteosarcomas appear in the long bones of the body, such as the femur (the thigh bone) the tibia (the shinbone) or the humerus (the bone that runs from the shoulder to the elbow), although they can appear in any bones.

2. What is the difference between osteosarcoma, which is a primary bone cancer and secondary bone cancers?

Osteosarcoma is a primary bone cancer. That means the cancer originates in the bone itself.

Secondary bone cancers are cancers that start in other parts of the body (such as the breast, prostate or lungs) and spread (metastasize) to the bone.

3. What groups of people are affected by osteosarcoma? What are the risk factors?

Osteosarcoma is most commonly found in adolescents and young adults. About 60% of cases occur in people between 10 and 20 years of age.

There are no known risk factors for osteosarcoma, though young people may be at higher risk if they suffer from Retinoblastoma or Li-Fraumeni syndrome (see glossary). Adults may be at higher risk if they have a history of Paget's disease or radiation therapy used to treat other cancers.

SYMPTOMS AND DIAGNOSTICS

4. What are the symptoms of osteosarcoma?

Symptoms of osteosarcoma may include:

- Persistent pain, swelling or a firm lump on a bone, especially on an arm or leg.
- A limp (if the tumor affects the leg).
- Pain or difficulty breathing (if the tumor affects the ribs).
- A bone fracture that occurs spontaneously or after a minor bump.

Other symptoms include gradually worsening pain and swelling in an arm or leg, near the knee or shoulder.

The pain from the tumor may occur when the patient is resting or may awaken the patient from a sound sleep.

5. How is osteosarcoma diagnosed?

After a full physical examination, the bone will be x-rayed. If the x-ray suggests osteosarcoma, further tests, such as a chest x-ray, blood test, CT scan, MRI, angiography, or bone scan may be performed. Finally, the diagnosis will be confirmed by a biopsy of the tumor.

TREATMENT

6. What is standard therapy for osteosarcoma?

The standard therapy for osteosarcoma is surgery with chemotherapy given before and after surgery. The chemotherapy is used to reduce tumor size or prevent any recurrence of cancer cells.

The percentage of destroyed tumor cells found in the tumor after surgery will indicate the type of chemotherapy treatment that should be used after the surgery.

7. What surgical procedures are used to treat osteosarcoma?

Surgery for osteosarcoma uses one of three different procedures: limb-sparing; rotational plasty and amputation. The development of limb-sparing surgery has led to a large decrease in amputation rates among people with osteosarcoma. The decision to try limb-sparing surgery depends upon the tumor size, location in the body and response to chemotherapy before the surgery.

Limb-sparing surgery removes the cancerous tumor and bone, replacing it with either a graft or prosthesis to make the limb as functional as possible. Seventy percent to 90 percent of osteosarcomas in the limbs can be treated by this method.

Rotation-plasty is a limb-sparing technique. The doctor removes a portion of the leg, including the knee. The lower part of the leg is rotated and reattached so that the ankle becomes the new knee, and a prosthetic device is attached to replace the ankle and foot.

Amputation is the removal of the limb. In most cases, prosthesis can be used to replace the limb.

PRIMARY BONE CANCERS

8. What are the other types of primary bone cancers?

The four main types of primary bone cancers are osteosarcoma, Ewing's sarcoma, chondrosarcoma and malignant fibrous histiocytoma.

Ewing's sarcoma, the second most common form of bone tumor in children and adolescents, is a very rare tumor whose risk factors are not yet known.

Chondrosarcoma, a tumor based in the cartilage, is the second most common type of tumor in primary bone cancers. Chondrosarcoma is usually found in adults over 30, with most patients between 50 and 70 years of age.

Malignant fibrous histiocytoma (MFH), the rarest of the four most common bone cancers, is primarily found in adults over 30. Although most cases develop for unknown reasons, a few have been linked to Paget's disease or radiation therapy used to treat other cancers.

GLOSSARY

Angiography, Chest x-ray, CT scan, MRI – Special tests done by a radiologist that give a more detailed view of bone, nearby muscles, blood vessels, organs and other tissues.

Biopsy – Removal of a small piece of tissue, which is then examined for cancer cells.

Bone scan – A test performed by a radiologist that allows the doctor to check the skeleton for the presence of cancer.

Cartilage – A dense, elastic tissue, which cushions bones at the joints, connects muscles with bones, and provides structure to the nose and other body parts.

Chondrosarcoma – second most common type of cancer to arise from bones, accounting for 20 to 25 percent of all primary bone cancers, most often found in adults.

Ewing's sarcoma – the second most common form of malignant bone tumor in children and adolescents.

Graft – replacement of diseased bone tissue with the patient's own tissue (autograft) or tissue from a donor (allograft).

Li-Fraumeni syndrome – (LFS) – a rare cancer syndrome that runs in families. LFS is caused by a gene mutation. Family members who have LFS are at risk for many different cancers, including breast cancer and osteosarcoma.

Limb-sparing surgery – The surgical removal of an arm or leg tumor so that the limb is preserved.

Malignant fibrous histiocytoma (MFH) – a type of cancerous tumor that can originate in either bone or, most often, in the soft tissues that connect, support or surround organs and other body structures.

Osteosarcoma – a primary cancerous tumor that develops in a bone.

Paget's disease of bone – a chronic skeletal disorder which may result in enlarged or deformed bones in one or more parts of the skeleton.

Primary cancer – Cancer can begin in any organ or tissue of the body. The original tumor is called the primary cancer or primary tumor. It is usually named for the part of the body or the type of cell in which it begins.

Prosthesis – a synthetic device used to replace a missing body part.

Retinoblastoma – a cancer in the retina of one or both eyes. It usually occurs in young children.

Rotation-plasty – a limb salvage technique. The doctor removes a portion of the leg, including the knee. The lower part of the leg is rotated and reattached so that the ankle becomes the new knee, and a prosthetic device is attached to replace the ankle and foot.

Secondary cancer – Cancer which has spread (metastasized) from another part of the body.

Syndrome -- a group of symptoms that indicate or characterize a disease, psychological disorder, or other abnormal condition.

Tumor – An abnormal mass of tissue. Tumors are either benign (non-cancerous) or malignant (cancerous).

The mission of The Bone and Cancer Foundation is to:

- Provide information to cancer patients and family member on the causes and current treatment of cancer that involves the bone.
- Provide information and serve as a resource for physicians, nurses, and other health professionals regarding the management of cancer that spreads to the bone.



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