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Paediatrics

# EWING'S SARCOMA: A REVIEW ARTICLE

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**ABSTRACT** Ewing sarcoma is the second most common bone cancer in children. It occurs most frequently in the long bones of the legs or arms, the pelvis, chest wall, spine and the skull, but can also begin in the soft tissues and not involve bone. This disease most often occurs in adolescents, with nearly half of cases arising between the ages of 10 and 20. Ewing sarcoma is somewhat more common in males than in females. The Ewing Family of Tumour" includes Ewing sarcoma, atypical Ewing sarcoma, and peripheral primitive neuroectodermal (PNET) of bone or soft tissue, and they are all treated in the same manner. A chromosomal mutation can be identified in the tumour cells of the Ewing Family of Tumour. The chromosomal mutation is not part of the genetic make-up of the person with the tumour, it is only in the tumour cells; the tumour is not inherited. The cause of the Ewing Family of Tumour is not yet known but it is believed this mutation plays an important role. Chemotherapy with alternating courses of 2 regimens: vincristine, doxorubicin, and cyclophosphamide; and ifosfamide and etoposide, Neutrophil support, Red blood cell and platelet support, Surgery and/or radiotherapy.

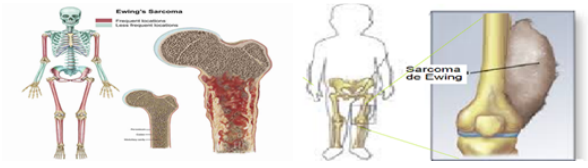
**KEYWORDS :**

## INTRODUCTION

The term sarcoma generally describes a cancer of connective tissues, including bone, muscle, cartilage, and other tissue types. Ewing sarcoma is a specific type of sarcoma in which the cancer cells are primitive and described as small round blue cells. Ewing sarcoma most commonly originates from a bone, but can also arise outside the bone, and is referred to as "extraosseous." Unlike osteosarcoma , it tends to affect bones of the axial skeleton about equally to the appendicular skeleton. Ewing sarcoma is the second most common cause of bone cancer in children.

## DEFINITION

A malignant tumor that arises in a primitive nerve cell within bone or soft tissue and affects children and adolescents, especially between ages 10 and 20.




## INCIDENCE

- Ewing's sarcoma is more common in males (1.6 male: 1 female) and usually presents in childhood or early adulthood, with a peak between 10 and 20 years of age.
- The incidence of these tumors peaks in the late teenage years. Overall, 27% of cases occur in the first decade of life, 64% of cases occur in the second decade, and 9% of cases occur in the third decade.

## CAUSES

- It's unclear what causes Ewing's sarcoma, as it doesn't appear to run in families. So far, research suggests that it's not related to exposure to radiation, chemicals, or other outside things in the environment.
- It appears that the cell DNA changes after birth, leading to Ewing's sarcoma. Why that happens remains unknown.
- One possibility is that it could be a second cancer in people who've been treated with radiation for another type of cancer

## TYPES

- ·**Bone tumor:** 87% of Ewing's sarcoma happens in the bone, often appearing in the thigh bones, pelvis, ribs, or shoulder blades. But the tumor can appear in any bone.
- **Soft tissue (extra-osseous) tumor:** This type of tumor affects the soft tissues around your bones, such as cartilage or muscle. It's rarely found in the soft tissue of the arms, legs, head, neck, chest, and abdomen.
- **Peripheral primitive neuroectodermal tumor (pPNET) :** This type of tumor is found in the nerves and can be detected in many parts of the body.
- **Askin tumor:** This is a type of pPNET tumor that's found in the chest.

## SIGNS AND SYMPTOMS

Signs and symptoms of Ewing sarcoma may include the following:

- Localized pain
- Back pain, which may indicate a paraspinal, retroperitoneal, or deep pelvic tumour
- Palpable mass
- Systemic symptoms of fever and weight loss, which often indicate metastatic disease
- Pain at the site of the mass, often with swelling
- Patients may have general symptoms such as loss of appetite, malaise, fatigue, and weight loss

**Examination for Ewing sarcoma includes the following:**

- Careful inspection and palpation of painful sites, as tumors of Ewing sarcoma can occur in almost any location; lesions of the long bones can present with a pathologic fracture
- Comprehensive neurologic examination to evaluate asymmetrical weakness, numbness, or pain; tumors close to bone can result in neuropathic pain
- Skin inspection for petechiae or purpura that may be caused by thrombocytopenia due to clinically significant bone marrow metastases
- Pulmonary auscultation for asymmetrical breath sounds, pleural signs, or rales that may indicate lung metastases

## DIAGNOSIS

No specific blood tests are diagnostic for Ewing sarcoma

- Complete blood count
- Blood cultures
- C-reactive protein levels
- Erythrocyte sedimentation rate
- Lactate dehydrogenase
- Cytogenetic and molecular studies
- Histology
- Imaging of the suspected primary lesion or of any region with symptoms
- Plain radiography in areas where a bony mass is palpated
- Magnetic resonance imaging.

## MANAGEMENT

Treatment for Ewing sarcoma includes the following:

- Chemotherapy with alternating courses of 2 regimens: (1) vincristine, doxorubicin, and cyclophosphamide; and (2) ifosfamide and etoposide
- Neutrophil support
- Red blood cell and platelet support
- Surgery and/or radiotherapy

## SURGICAL MANAGEMENT

- **Amputation**
- **Limb Salvage Surgery for Localized Ewing's Sarcoma:**
- **Surgery for Metastatic or Recurrent Ewing's Sarcoma**
- **Reconstructive Surgery:** These include bone grafts (using the patient's own bone or bone from a donor) and prosthetic implants.
- Rotationplasty: is a technique used commonly in patients with Ewing's sarcoma that involves the lower femur or upper tibia.

## CONCLUSION

Ewing sarcoma can spread to other parts of the body quite quickly. The earlier its diagnosed, the better chance there is of the treatment being successful. The cancer can also come back after treatment, so regular check- ups will be offered to look for any signs of recurrence.

## REFERENCES

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