Ewing's Sarcoma Family of Tumors:
An Overview From Diagnosis to Survivorship

Meredith Lahl, RN, MSN, CNS, CPON®,
Vicki L. Fisher, RN, CNP, CPON®, and Kelly Laschinger, RN, MSN, CPNP

The Ewing's sarcoma family of tumors (ESFT) is a malignant primary bone tumor often involving soft tissue that affects not only children but also young adults. Since 1992, with the addition of ifosfamide and etoposide to standard chemotherapy for primary tumors, much improvement has been made in the treatment of ESFT, with a primary focus on children. Though often recognized as a childhood cancer, it can affect individuals into the middle years of their lives, but little is known about the outcomes of adults with ESFT. ESFT, which includes Ewing's sarcoma, extraosseous Ewing's sarcoma, Askin tumor, and primitive neuroectodermal tumor, is the second most common primary malignant bone tumor in children and adolescents. It accounts for 10% of primary malignant bone tumors in children and 3% of all childhood malignancies. The most common presenting symptoms of ESFT are pain or swelling. Treatment for ESFT consists of a multimodal approach, including chemotherapy, radiation therapy, and surgery. Children and young adults with Ewing's sarcoma face many physical challenges from their illness and the complications of their treatments. Nurses play an instrumental role in assessment techniques, which lead to prompt evaluation and intervention. Nurses are vital in the education and reinforcement of supportive care needs for this patient population.

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The Ewing's sarcoma family of tumors (ESFT) is a malignant primary bone tumor often involving soft tissue; it affects children and young adults. It was first described in 1921 by James Ewing as a tumor of the shaft of long bones, which, unlike osteosarcoma, was radiosensitive (Pizzo & Poplack, 2005). Since 1992, with the addition of ifosfamide and etoposide to standard chemotherapy for primary tumors, much improvement has been made in the treatment of ESFT, with a primary focus on children. The improvement stems from the initial speculation that it was a tumor of endothelial origin; recent evidence shows a neural origin (Pizzo & Poplack, 2005). Though primarily recognized as a childhood cancer, it can affect individuals into the middle years of their lives, but little is known about the outcomes of adults with ESFT.

At a Glance

- Ewing's sarcoma family of tumors (ESFT) is a malignant primary bone tumor that affects children and young adults.
- Treatment for ESFT involves a combination of chemotherapy, radiation, and surgery.
- Because of the aggressive treatment, patients experience many toxicities and side effects.

Incidence and Epidemiology

ESFT, which includes Ewing's sarcoma, extraosseous Ewing's sarcoma, Askin tumor, and primitive neuroectodermal tumor, is the second most common primary malignant bone tumor in children and adolescents. It accounts for 10% of primary malignant bone tumors in children.

The tumors occur most commonly in the second decade of life (Pizzo & Poplack, 2005), and approximately 80% of those with Ewing's sarcoma are younger than 20 (Lanzkowsky, 2005). ESFT has been reported in those older than 30 and the very young, but it is uncommon (Pizzo & Poplack). ESFT shows a slight male predominance.

A striking difference exists in the incidence of ESFT among racial backgrounds. The incidence is very rare (fewer than 2% of cases reported) in the black population (Horowitz, Tsokos, & DeLaney, 1992). ESFT has been reported in India and Japan but is distinctly uncommon in China (Pizzo & Poplack, 2005). The incidence of the tumors in the United States is 2.1 cases per million children (Grier, 1997; Pizzo & Poplack).

Meredith Lahl, RN, MSN, CNS, CPON®, is a pediatric clinical nurse specialist at the Cleveland Clinic in Ohio; Vicki L. Fisher, RN, CNP, CPON®, is a program manager of the pediatric bone marrow transplant program at the University Hospitals of Cleveland in Ohio; and Kelly Laschinger, RN, MSN, CPNP, is a pediatric nurse practitioner at University Hospitals of Cleveland. No financial relationships to disclose. (Submitted February 2007. Accepted for publication October 26, 2007.)

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