

## INTRODUCTION

One of the most difficult aspects of illness in children is the immobility it often imposes on children. Children's natural tendency to be active influences all aspects of their growth and development. Impaired mobility presents a challenge to children, their families, and their caregivers (*Helman, 2011*).

Bone tumors account for about 6% of all malignant neoplasms in children. Approximately 90% of all primary malignant bone tumors in children are either osteogenic sarcoma or Ewing sarcoma; osteosarcoma, the most common, occurs in 56% of all cases. The peak age for pediatric bone tumors is 15 years, and they occur more often in boys (*Holmes et al., 2005*).

The incidence of osteosarcoma increases steadily with age; a relatively dramatic increase in adolescence corresponds with the growth spurt. Osteosarcoma is rarely diagnosed in children younger than 5 years (about 1% of cases). Children diagnosed during their growth spurt are taller than average, while patients identified in adulthood have an average height (*Kager et al., 2010*).

Children have different needs than adults. They often require orthopedic care administered by specially trained experts in the care of children. That care is best provided in a facility designed exclusively for children. It includes pediatric orthopedic surgeons, pediatric radiologists, and pediatric anesthesiologists who specialize exclusively in the care of

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children. This creates an environment that is comfortable and safe for the child (*Rodts, 2004*).

The nurse plays a vital role in caring for patients with osteosarcoma. From the very outset when the disease is explained to children and their family, the nurse provides comfort and support, as well as enhancing and explaining the information provided by the physician. This communication will provide a perspective of the role assumed by the nurse in effort to ensure total care of children and their families (*Smith et al., 2010*).

An orthopedic nurse is a specialty nurse trained in orthopedic problems such as fractures and is an expert in neurovascular status monitoring, traction, casting and continuous motion therapy. Nurses' skills, interventions, attitudes, communication and continuity of care constitute the essential components of orthopedic nurse care (*Watters and Moran, 2006*).

Attitudes of nurses caring for orthopedic patients affect the quality of care provided. A recent research on positive and negative attitudes of such nurses has shown that knowledge deficits shape most of their negative attitudes. The cultural background of nurses also has an influence on the attitudes and there are reports of nurses' disagreements with patient's self-report, especially in pain assessment (*Harper et al., 2007*).

Nurses are best placed to work with the osteosarcoma children admitted to hospital by developing close relationships with these

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children and their families, they need to liaise closely with the specialist team and share information to ensure that children and their families receive optimal care during hospitalization and after their discharge home (*Valentine and Lowes, 2007*).

Educational programs such as activities start by diagnosing the assessment program's goals and objectives, and the methods of teaching and program evaluation. They involve a mix of objectives based on hospital policies and rules. A program also outlines the specific steps to be taken to achieve its objectives along with the time, money, and human resources required to complete it. Educational program in nursing is very important to increase the quality of nursing education and clinical practice (*Barron's Finance, 2006*).

## AIM OF THE STUDY

**The present study aimed to:** Enhancing the perioperative care provided by nurses for children with osteosarcoma and their families.

**Through:**

1. Assessing the nurses' knowledge and practices related to perioperative care provided to the children with osteosarcoma and their families.
2. Developing and disseminating a handout for nurses about perioperative care.
3. Evaluating the effects of the educational program among nurses about care given to the children with osteosarcoma, perioperatively.

**Study questions:**

- 1- Does an educational program have a positive effect on nurses' knowledge and practices?
- 2- To what extent can an educational program affect nurses' performance to ward perioperative care for children?

## OSTEOSARCOMA

**Osteosarcoma** is known as osteogenic sarcoma. Sarcoma is a type of cancer that starts in the cells of soft tissues or bone. Osteosarcoma osteogenic sarcoma, is a cancer that makes abnormal bone, it usually starts within the bone and rarely begins in muscles. Most common in children and adolescents, it occurs commonly in the bones around the knee. The arm bone near the shoulder is the second most common place for this cancer to start, but it can start in other bones as well. Like other cancers, osteosarcoma can spread into nearby tissues. It can also spread to the lungs and to the other bones. Rarely, it can spread into other organs (*Gebhart et al., 2008*).

The cells that form osteosarcoma make the bone matrix, the material that makes bones strong. But, the bone matrix produced by osteosarcoma cells is not as strong as the bone matrix produced by normal bone cells (*Malawer et al., 2011*).

The incidence of osteosarcoma seems to peak at the same time that adolescents are growing most rapidly. Osteosarcoma is more common among children who are taller or grow more rapidly (*Gorlick et al., 2011*).

Osteosarcoma is an extremely malignant primary cancer of long bones. Evidence of malignant osteoid bone and/or cartilage formation with destructive lesions and sclerosis is characteristic of the disease. Differentiating osteosarcoma from other tumors is defined by the production of extensive

incompletely mineralized matrix that is seen with histological staining (*Yasko et al., 2010*).

Treatment involves surgery, usually followed by chemotherapy or radiation. The site of the tumor is the most important prognostic factor, because it determines whether the tumor can be surgically removed or not (*National Comprehensive Cancer Network, 2012*).

**Epidemiology:**

Osteosarcoma is derived from the primitive bone-forming mesenchymal cells and is the most common primary bone malignancy. The incidence rates and the 95% confidence intervals of osteosarcoma for all races and both sexes are 4.0 (3.5–4.6) for the range 0–14 years and 5.0 (4.6–5.6) for the range 0–19 years per year per million persons. The incidence of osteosarcoma has always been considered to be higher in males than in females, occurring at a rate of 5.4 per million persons per year in males versus a rate of 4.0 per million in females (*Lewis et al., 2007*).

**Sites of osteosarcoma:**

Osteosarcoma commonly occurs in the long bones of extremities near the metaphyseal growth plates. The most common sites are the femur (42%, with 75% of tumors in the distal femur) the tibia (19%, with 80% of tumors in the proximal tibia), and the humerus (10%, with 90% of tumors in the proximal humerus). Other likely locations are the skull or

jaw (8%) and the pelvis (8%). Cancer deaths due to bone and joint malignant neoplasms represent 8.9% of all childhood and adolescent cancer deaths. Death rates for osteosarcoma have been declining by about 1.3% per year (*Wang et al., 2012*).

**Incidence of osteosarcoma:**

Osteosarcoma is the eighth most common form of childhood cancer, comprising 2.4% of all malignancies in pediatric patients and approximately 20% of all primary bone cancers. Incidence rates for osteosarcoma in U.S. patients under 2 years of age are estimated at 5.0 per million per year in the general population, with a slight variation between individuals (*Steliarova, 2004*).

Osteosarcoma represents approximately 55% of child and adolescent malignant bone tumors in the US. It is rarely diagnosed before the age of five, but the incidence increases with age until around puberty (*Kager et al., 2010*).

In Egypt according to the El-Gharbia Cancer Registry (ECR), department of statistics, osteosarcoma accounts for 4 per million of all childhood malignancies, and accounts for 4% of all childhood cancer according to Children Cancer Hospital Egypt (CCH) (*El-Gharbia Cancer Registry, 2007*).

Most osteosarcomas are not caused by inherited genetic mutations. Rather, they are a result of mutations acquired during a person's lifetime. People with these mutations do not pass them on to their children (*Wang et al., 2012*).

## **Clinical Presentation**

### **Symptoms:**

The symptoms of bone cancer depends on the size and location of the tumor, pain is the most common symptom. Tumors arising in or around the joints often cause swelling and tenderness. The tumors can also weaken the bones and interfere with normal movement, that causing fractures. Other symptoms can include weight loss, fatigue and anemia (too few red blood cells) (*Bielack and Carrel, 2008*).

The most common symptoms of osteosarcoma are pain and swelling in a child's leg or arm. It occurs most often in the longer bones of the body such as above or below the knee and in the upper arm near the shoulder. Pain may be worse during exercise, at night and a lump or swelling may develop in the affected area up to several weeks after the pain starts. Pain that persistently wakes the child up at night or pain at rest is of particular concern. In osteosarcoma of the leg, the child may also develop an unexplained limp. In some cases, the first sign of the disease is a broken arm or leg, because the cancer has weakened the bone to make it vulnerable to a break (*Lewis et al., 2007*).

### **Other symptoms of osteosarcoma include:**

Joint tenderness or inflammation, fractures due to bone weakness, limited range of motion and non-specific symptoms like fever, unintentional weight loss, fatigue, and anemia can also be symptoms of osteosarcoma, but they are also indicators



of other less severe condition (*American Cancer Society, 2008*).



**Figure (1):** A clinical photograph of a large mass of the distal femur (*National Comprehensive Cancer Network "NCCN", 2012*). Clinical Practice Guidelines in Oncology: Bone Cancer. Version 2. Available at: [http://www.nccn.org/professionals/physician\\_gls/pdf/bone.pdf](http://www.nccn.org/professionals/physician_gls/pdf/bone.pdf). Accessed November 16.

Swelling in this area is the next most common symptom, although it may not occur until several weeks after the pain starts (*Gebhart et al., 2008*).

### **Types:**

There are different types of osteosarcoma based on how they look when their cells are examined closely, depending on how they look under the microscope (basically, how much they look like normal bone cells) osteosarcomas are classified into different grades with each grade telling how likely will the cancer spread to other parts of the body (*Lerner and Antman, 2011*).

**Low grade** - The tumor has few dividing cells. These tumors can often be completely removed with surgery and do not require chemotherapy (*Lerner and Antman, 2011*).

**Intermediate grade** - These tumors are categorized as periosteal, or formed from the dense membrane that wraps the bone (*Lerner and Antman, 2011*).

**High grade** - The tumor has many dividing cells and many dead cells. These tumors tend to be aggressive and usually treated with both surgery and chemotherapy. Most osteosarcomas in children are high grade (*Bielack and Carrle, 2008*).

**Staging:**

The 'stage' of a cancer is a term used to describe its size and whether it has spread beyond its original site. Knowing the particular type and stage of the cancer helps to decide on the most appropriate treatment, most patients are grouped depending on whether cancer is found in only one part of the body (localized disease) or whether the cancer has spread from one part of the body to another (metastatic disease) (*European Sarcoma Network Group: ESMO, 2012*), it can be classified into the following stages:

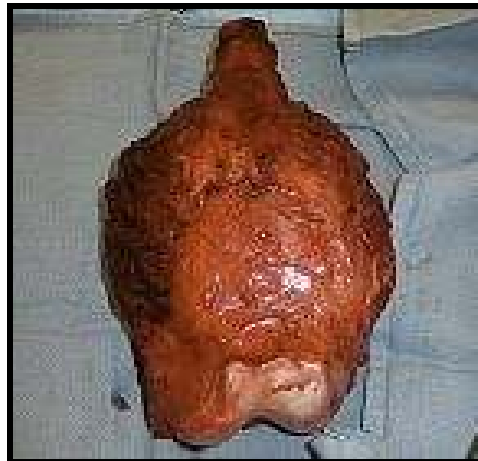
**Stage 1A:** The cancer is low-grade and is only found within the hard coating of the bone.

**Stage 1B:** The cancer is low-grade, extending outside the bone and into the soft tissue spaces that contain nerves and blood vessels.

**Stage 2A:** the cancer is high-grade and is completely contained within the hard coating of the bone.

**Stage 2B:** The cancer is high-grade and has spread outside the bone and into surrounding soft tissue spaces that contain nerves and blood vessels (*European Society of Medical Oncology (ESMO), 2010*).

**Stage 3:** The cancer can be low- or high-grade and is either found within the bone or extends outside the bone. The cancer has spread to other parts of the body or to other bones not directly connected to the bone where the tumor started. If the cancer comes back after initial treatment, this is known as recurrent or relapsed cancer (*Koshkina and Corey, 2008*).



**Figure (2):** Photograph of osteosarcoma at the distal femur (*European Society of Medical Oncology "ESMO", 2010*). Clinical practice guideline on diagnosis, treatment, and follow-up of bone sarcomas. Annals of Oncology, Volume 21: Supplement 5.

### **Diagnosis:**

The diagnosis of osteosarcoma requires a detailed medical history, physical examination and X-rays to detect any changes in bone structure. **Magnetic Resonance Imaging (MRI)** scan of the affected area will find the best area to biopsy and show whether the osteosarcoma has spread from the bone into nearby muscles and fat. The bone biopsy is to obtain a sample of the tumor for examination in the lab, sometimes achieved by a needle biopsy, using a long hollow needle to take a sample of the tumor. Alternatively, an open biopsy may be ordered. If diagnosis of osteosarcoma is made, CT chest scans as well as a bone scan and sometimes additional MRI studies may be required. These will show if the cancer has spread to any part of the body beyond the original tumor (*Geller and Gorlick, 2010*).



**Figure (3): AP and Lateral X-Rays of the mass. (*National Comprehensive Cancer Network "NCCN", 2011*). Clinical Practice Guidelines in Oncology: Bone Cancer. Version 2. Available at: [http://www.nccn.org/professionals/physician\\_gls/pdf/bone.pdf](http://www.nccn.org/professionals/physician_gls/pdf/bone.pdf). Accessed November 16.** This x-ray shows a malignant bone tumor (osteogenic sarcoma) of the knee. This type of tumor is usually seen in adolescents (around 15 years old). This tumor extends from the bone into the surrounding tissue.



**Figure (4): T1-weighted coronal MRI image shows a large distal femur** (*European Society of Medical Oncology, "ESMO", 2010*). Clinical practice guideline on diagnosis, treatment, and follow-up of bone sarcomas. *Annals of Oncology*, Volume 21: Supplement 5.

#### **Treatment of osteosarcoma:**

Treatment for osteosarcoma may involve a combination of therapies including surgery, chemotherapy and rarely, radiation. In most cases, children receive chemotherapy before surgery (neoadjuvant), a surgical procedure to remove the tumor and additional chemotherapy after surgery (adjuvant) (*Paiva et al., 2005*).

#### **Chemotherapy:**

Chemotherapy is a group of drugs that interfere with the cancer cells' ability to grow or reproduce. Different groups of chemotherapy drugs work in different ways to fight cancer cells and shrink tumors. Often, a combination of chemotherapy drugs is used. Certain chemotherapy drugs may be given in a

specific order, depending on the type of cancer it is being used to treat (*Myers et al., 2008*).

Chemotherapy is a systemic treatment, introduced to the bloodstream and travelling throughout the body to kill cancer cells. The chemotherapy used for osteosarcoma is given intravenously. Usually, several drugs are given together (*Chou et al., 2007*).

**Systemic therapy:**

***Polychemotherapy:***

Currently, doxorubicin, cisplatin, high-dose methotrexate with leukovorin-rescue and ifosfamide are considered the most active agents against osteosarcoma, but the ideal combination remains to be defined (*Goorin et al., 2003*).

Most current protocols include a period of preoperative (neoadjuvant) chemotherapy, even though this has not proved to add a survival benefit over postoperative (adjuvant) chemotherapy alone. The extent of histological response to preoperative chemotherapy, however, offers important prognostic information. Current prospective trials evaluate whether altering postoperative chemotherapy in poor responders improves outcomes. The use of high-dose chemotherapy followed by retransfusion of autologous hematological stem cells has not led to improved outcomes (*Meyers et al., 2008*).

**Treatment protocols for osteosarcoma:**

First-line treatment consists of doxorubicin and cisplatin; high-dose methotrexate, cisplatin, ifosfamide, and etoposide, cisplatin, and epirubicin. Doxorubicin and cisplatin therapy, doxorubicin ( $25 \text{ mg/m}^2$  IV) on days 1-3 plus cisplatin ( $100 \text{ mg/m}^2$  IV) on day 1; repeat cycle every 21 days. MAP (high-dose methotrexate, cisplatin, and doxorubicin), neoadjuvant setting: high-dose methotrexate ( $12 \text{ g/m}^2$  IV) given over 4 hours on weeks 0, 1, 5, 6, 13, 14, 18, 19, 23, 24, 37, and 38, alternating with cisplatin ( $60 \text{ mg/m}^2$  IV) plus doxorubicin ( $37.5 \text{ mg/m}^2/\text{day}$  IV) for 2 days each on weeks 2, 7, 25, and 28 (*Goorin et al., 2003*).

Adjuvant setting: high-dose methotrexate ( $12 \text{ g/m}^2$  IV) given over 4 hours on weeks 3, 4, 8, 9, 13, 14, 18, 19, 23, 24, 37, and 38, alternating with cisplatin ( $60 \text{ mg/m}^2$  IV) plus doxorubicin ( $37.5 \text{ mg/m}^2/\text{day}$  IV) for 2 days on weeks 5, 10, 25, and 28; 2 cycles are given preoperatively, and 4 cycles are usually given postoperatively, they require administration of 15 mg leucovorin every 6 hours for 10 doses, starting 24 hours after initiation of high-dose methotrexate. If methotrexate elimination is delayed, then immediately administer 50 mg IV leucovorin every 3 hours until serum methotrexate levels are undetectable. Doxorubicin, cisplatin, ifosfamide, and high-dose methotrexate: ifosfamide ( $15 \text{ g/m}^2$ ) plus methotrexate ( $12 \text{ g/m}^2$ ) plus cisplatin ( $120 \text{ mg/m}^2$ ) plus doxorubicin ( $75 \text{ mg/m}^2$ ) (*Ferrari et al., 2005*).