# Features of Bone Sarcomas at MAHAK Children Hospital, Tehran, Iran during 2007-2009

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## Abstract

**Background:** Bone sarcomas are the most common malignancies of bone tissues in children, and are classified into two groups as osteosarcoma and Ewing's Sarcoma. Treatment and prognosis depend on the subtype and grade of the tumor. The goal of this study was to evaluate the features bone sarcoma in patients referred to MAHAK children hospital since 2007 to 2009.

**Materials and Methods:** This was a retrospective study of patients with bone sarcoma referred to MAHAK children hospital for continuing their treatment or as a new case of cancer. All of the patients had local pain, local swelling, and decreased range of motion and pathologic fractures as the presenting symptom. All patients underwent bone scan, chest computed tomography (CT) scan, chest X- ray, echocardiography, hearing tests, and bone marrow aspiration/biopsy. All of the data were analyzed by SAS software.

Results: In this study, 36 patients with bone sarcoma were included with the age range of 3 to 20 years old (55% male and 45% female). Analysis showed that local pain and local swelling were the most frequent local manifestations in patients. The most common primary tumor site was distal femur (30%). Treatment for the patients at MAHAK children hospital is performed according to German protocol. Out of 36 patients, 75% were followed until 2008 and 25% until 2009.

**Conclusion:** According to the analysis we can conclude that these data are similar to other studies. To achieve the best results, oncologists have to modify the treatment of patients completely.

**Keywords:** Sarcoma, Manifestations, Protocol.

## Introduction

Sarcomas originate in bone, muscle, fibrous tissue, blood vessels, fat tissue, and some other tissues.[1] They are divided into malignant (osteosarcoma, chondrosarcoma, Ewing's tumor, malignant fibrous histiocytoma, fibrosarcoma, and chordoma) and benign bone tumors.[2] Osteosarcoma and Ewing's sarcoma are the most common malignancies of bone tissues in children.[3] Most bone sarcomas in younger people affect the leg, while in adults there are more variable sites.[4] Several bone tumors have clinical manifestations similar to osteosarcoma.[5] In Ewing's tumor, irradiation has almost always given enough control of the local disease to make amputation unnecessary.[6] Giant cell tumors, if surgically inoperable, can be cured by radiotherapy, but most would agree that adequate surgery is the treatment of choice whenever possible.[7] Chondrosarcomas may be clinically very similar to osteosarcomas, and can also be treated using the same treatment regimen (irradiation and delayed amputation).8 Follow-up care varies depending on the type and grade of the tumor.[9]

The goal of this study was to evaluate the frequency of bone sarcomas at MAHAK children hospital from 2007 to 2009 by pathological and clinical examinations..

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## Materials and Methods

### *Patients*

The study group consisted of patients who have been referred to MAHAK children hospital from 2007 to 2008. Some of them were diagnosed in other centers and then referred to MAHAK for continuing their treatment, and others were diagnosed for the first time at MAHAK hospital.

### *Clinical manifestations*

We evaluated all patients regarding local pain, local swelling, decreased range of motion, and pathologic fractures. Also we performed bone scan, chest CT scan, chest X-ray, echocardiography, hearing tests, and bone marrow.

### *Diagnosis and treatment*

According to clinical manifestations and primary tumor site, we classified patients. The treatment of childhood bone sarcoma in MAHAK children hospital is performed according to German protocol. Initially each patient received five courses of chemotherapy using Adriamycin, Cisplatin, or Methotrexate; then the patient underwent surgery (either limb salvage or amputation), and finally seven courses of chemotherapy by Adriamycin, Cisplatin, or Methotrexate were administered (figure 1).

### *Analysis*

Data was analyzed using Epi-info and SAS software.

## Results

Thirty six patients with bone sarcoma were evaluated in this study, of whom twelve patients had been diagnosed in other centers during 2004-2006 and referred to MAHAK hospital. Table 1 shows the frequency in each year of the study.

Out of 36 patients, there were 20 male (55%) and 16 female (45%). The age range at diagnosis was 3 to 20 years old. We categorized them into four age groups; 0-5 years old, 6-10 years old, 11-15 years old, and more than 16 years old. Most patients belonged to the third group (11-15 years old) (table 2). In each group the frequency of male patients was the same as females except the third group in which males were nearly 1.5 times more than females (table 3).

Analysis showed that out of 36 patients, 80% had local pain, 30% local swelling, 6% decrease range of

**Table 1.**

| Year | Frequency | Percent |
|---|---|---|
| 2004 | 2 | 5% |
| 2005 | 5 | 15% |
| 2006 | 5 | 15% |
| 2007 | 14 | 44% |
| 2008 | 10 | 21% |

**Table 1. Diagnosed/referred patients in each year.**

**Table 2.**

| Age groups (years) | Frequency | Percent |
|---|---|---|
| 0 - 5 | 4 | 11% |
| 6 - 10 | 4 | 11% |
| 11 - 15 | 21 | 58% |
| More than 16 | 7 | 20% |
| **Total** | **36** | **100%** |

**Table 2. The age at diagnosis groups.**



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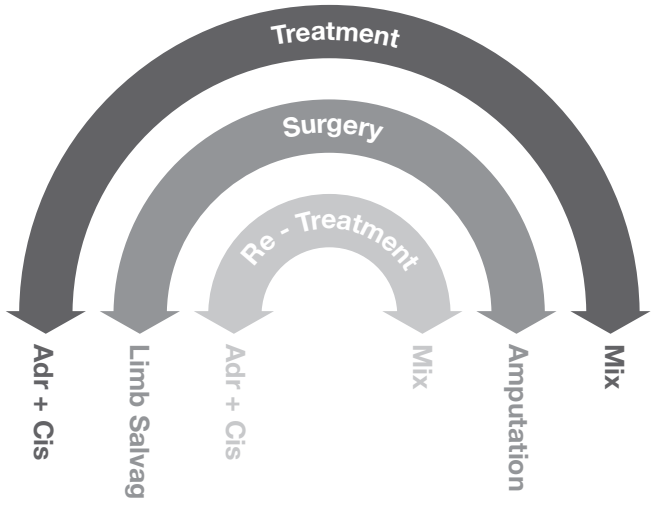
**Figure 1.** Treatment plan.




motion, and 6% pathologic fractures. There were 16 patients with abnormal bone scan and 2 patients with abnormal chest CT scan.

The primary tumor site was categorized as distal and proximal femur, distal and proximal fibula, distal and proximal tibia, distal and proximal humorous, pelvis, and others (chest, knee, mandible, sacral, and sacrum). Table 4 shows the frequency of primary tumor site. We classified patients in five diagnostic groups (figure 2); Classic (osteoblastic, Chondroblastic, fibroblastic), telangiectatic, small round cell tumor, periosteal, and parosteal.There were three sections of treatment for considered patients. Analysis showed that 16 patients (44%) received adriamycin+cisplatin and 20 (56%) received methotrexate at the first section of their treatment. Then 16 patients (44%) had limb salvage, 6 (17%) had amputation and 14 (39%) did not accept to undergo any surgery in the second section of their treatment. At last, 11 (30%) of patients were cured with adriamycin+cisplatin, 6 (17%) received methotrexate, and 53% still are continuing their last section of treatment. Out of 36 patients who were enrolled in this study, 8 patients (22%) died and 28 (78%) are alive. 25% of patients who are alive are still under follow up examinations.

**Figure 2**. The frequency of classified diagnostic groups.

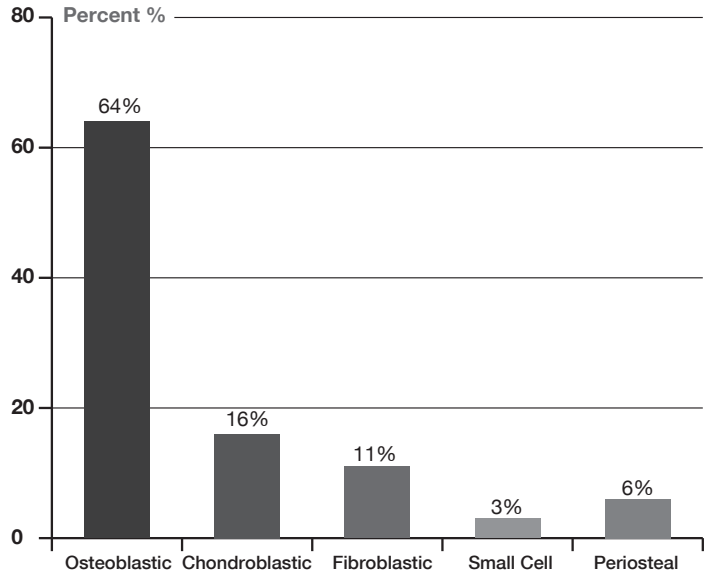



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**Table 3.**

| Age groups (years) | Male No (%) | Female No (%) |
|---|---|---|
| 0 - 5 | 2 (50%) | 2 (50%) |
| 6 - 10 | 2 (50%) | 2 (50%) |
| 11 - 15 | 13 (62%) | 8 (38%) |
| More than 16 | 3 (43%) | 4 (57%) |

**Table 3. The frequency of genders in the age at diagnosis groups.**

## Discussion

Sarcoma is a cancer of the soft tissue (muscle, fat, nerve, and connective tissue) or bone. Sarcomas encompass a group of over 40 different types of tumors. Osteosarcoma is one of the most common primary bone cancers. It accounts for about 5% of all childhood cancers. It most often occurs in young people between the age of 10 and 30, but about 10% of osteosarcoma patients develop in people in their 60s and 70s. It is rare during middle age, and is more common in males than females. These tumors develop most often in bones of the arms, legs, or pelvis. Treatment and prognosis depends on the subtype and grade of the tumor. A five year event free survival could be expected in 85% of patients. High grade tumors consist of immature cells which require aggressive chemotherapy, 75 percent of whom have a five year survival.

Chondrosarcoma is the second most common primary bone cancer. This cancer is rare in people younger than 20 years. After age 20, the risk of getting a chondrosarcoma increases until about age 75 years. Women get this cancer as often as men. Chondrosarcomas can develop in bones such as pelvis, leg, and arm. Occasionally, chondrosarcoma will develop in the trachea, larynx, and chest wall. Other sites are the scapula (shoulder blade), ribs, and skull.

Ewing's tumor is the third most common primary bone cancer, and the second most common in children, adolescents, and young adults. The most common sites for this cancer are pelvis, chest wall (such as the ribs or shoulder blades), and long bones of legs or arms. This cancer is most commonly seen in children and teenagers and is rare in adults over age 30 years. Ewing tumors occur most often in white people and are rare among African Americans and Asian Americans. It occurs in 0.6 per million people and is most common in the second decade of life. Fifty percent of patients without metastases may have a long term disease free survival.

According to different literature and reviews, we can conclude that survival rate and follow up of childhood bone sarcomas is similar to other countries. It could not be overemphasized that for gaining the best results of follow up oncologists have to modify the treatment of patients completely.

**Table 4.**

| Site | Frequency | Percent |
|---|---|---|
| Distal femur | 11 | 30% |
| Proximal femur | 5 | 25% |
| Proximal fibula | 3 | 8% |
| Distal tibia | 1 | 3% |
| Proximal tibia | 1 | 3% |
| Distal humorous | 4 | 19% |
| Proximal humorous | 1 | 3% |
| Pelvis | 2 | 6% |
| Other | 15 | 41% |
| **Total No. of patients** | **36** | **100%** |

**Table 4. The frequency of primary tumor sites.**

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