**Osteosarcoma in Adolescents**

**Introduction**

Osteosarcoma (OS) is the most commonly occurring bone tumor.1 It typically occurs in either bones around the knee or in the proximal arm.1 It accounts for approximately 10% of all sarcomas.2  In the United States, approximately 400 new cases are diagnosed annually.1  The peak incidence is during the second decade of life during the adolescent growth spurt.1  The incidence is higher in black children compared to Caucasians and it is more likely to affect males than females.1 This paper will focus on treatment of OS affecting the knee.

**Etiology and Pathology**

Osteosarcoma is theorized to result from an aberrant process of normal growth and remodeling. Growth factors have been suggested as important in the pathogenesis of OS.3 Having a tall stature or an early pubertal growth spurt have been associated as an etiology of OS.3  Osteosarcoma most frequently arises in the metaphysis of the most quickly growing bones during adolescence.1,4

Two suppressor genes, p53 and Rb, have significant roles in the tumorigenesis of OS.1  A genetic predisposition is also present in patients with hereditary retinoblastoma.1  Osteosarcoma has been associated with hereditary cancer syndromes.2 It is a primary bone tumor derived from bone-forming mesenchyme in which “the malignant proliferating spindle cell stroma produce osteoid tissue or immature bone”. 4 The resultant tumor is highly malignant. Survival rates are currently 75% for localized OS and 30% for patients with metastatic disease.1 Common sites of metastasis include the lungs and other bones.1 An adolescent being treated for OS is at risk for numerous impairments including fatigue, decreased ROM, decreased strength, poor motor control, pain, swelling, leg length difference, gait abnormalities, inability to perform activities of daily living, decreased functional mobility and fracture.5,6

**Clinical Manifestations**

***Presentation and Diagnosis***

Initial diagnosis may be made from a compilation of symptoms including bone pain which may be worse with weight bearing, limited range of motion (ROM), a limp, tenderness, redness or swelling around the tumor site.7,8  Pain may also be worse at night.7  Some children present with a pathological fracture.1  Medical work up to confirm the diagnosis includes blood tests, plain film x-rays, magnetic resonance imaging of the bone, computerized tomography of the chest, radionuclide bone scan and bone biopsy.1

***Medical Treatment and Surgical Intervention***

Treatment of OS is two-fold and includes chemotherapy and surgery. Osteosarcoma is chemosensitive and treated with multiple agents including cisplatin, doxorubicin, high dose methotrexate and ifosfamide.1  Three options are available for surgical treatment of OS of the knee. Determinants of which procedure is most appropriate is multifactorial and may include the child’s age, tumor location, vascular and nerve involvement, and family preference.

Traditional treatment of OS involves limb amputation.4  Children often adapt well to this option and have excellent functional outcomes.9  Many advances in prosthetic devices make this a viable option. A second option is rotationplasty where the distal femur and proximal tibia are resected and the foot rotated 180 degrees with the residual lower limb then reattached to the femur.4,9,10  The foot then serves as a knee joint.4,9,10

Lastly, limb sparing procedure is possible in 95% of cases of localized OS.1,9 During limb sparing, bony tumor is removed and the limb is reconstructed with an endoprosthesis.4,11  There is bone and soft tissue loss with this procedure.4,11  It may not be beneficial for a younger child secondary to resultant leg length difference with growth.4

**Physical Therapy Interventions**

The Guide to Physical Therapist Practice can assist in guiding evaluation and treatment of patients undergoing medical management and treatment of OS. During the acute, inpatient phase practice patterns including 4C: Impaired Muscle Function and 6B: Impaired Aerobic Capacity/Endurance Associated With Deconditioning may be appropriate. Many treatment interventions for patients with OS are core physical therapy skills.

The American College of Sports Medicine (ACSM) has developed guidelines for exercise in cancer survivors.12  They recommend exercise prescriptions should be individualized and based on pretreatment “aerobic fitness medical comorbidities, response to treatment, and the immediate or persistent negative effects of treatment that are experienced at any given time.”12

One treatment intervention during acute hospitalization could include decreasing fatigue, a common problem for children receiving chemotherapy. As the children are often immuno-compromised and must remain in their rooms, a portable bike, treadmill or step can be brought to them for exercising. A 2007 study demonstrated decreased fatigue when children exercised for 30 minutes twice daily for 2 to 4 days of hospitalization.13

Following surgery, depending on the procedure performed, 3 guide patterns may be applicable including: 4H, I or J: Impaired Joint Mobility, Motor Function, and Range of Motion associated with arthroplasty, bony or soft tissue surgery, and amputation respectively.14

An important physical therapy intervention following limb sparing surgery is knee range of motion (ROM) exercises. Marchese et al demonstrated that decreased knee ROM following limb sparing surgery affected functional mobility and quality of life.5  Carty and colleagues demonstrated that quadriceps strength and knee flexion ROM were associated with impairment and disability and these areas should be a focus of post-operative rehabilitation.11  Hip and knee ROM are important for functional activities such as walking and stair climbing. Range of motion and quadriceps strengthening can be achieved by sit to stand from different height benches while shooting a basketball into a hoop in the clinic. This would be an activity that an active, athletic teenage male may enjoy.

A third PT intervention following either amputation or rotationplasty, where a prosthesis will be worn, should include functional training with the prosthesis. Following training by a physical therapist including obstacle management, strengthening by climbing boxes and ladders, and functional training such as carrying water, subjects showed improved walking, physiological cost and maximal weight bearing on the amputated lower extremity.15  Instruction in running should be performed so that the child can return to age appropriate play and sports with peers.4 These activities could be practiced in the outpatient clinic on indoor and outdoor terrain.

**Community Resources and Activities**

Cancer is often a very emotionally charged diagnosis for families. For families facing a diagnosis of OS, several informational and support resources are available. On a national level, organizations such as the American Cancer Society and National Cancer Institute are resources available to provide information and support for families.16,17

Caring Bridge is a nonprofit organization that allows people to keep in touch by sending and receiving messages in one place.18  Families use this to update family, friends and staff. St Jude’s Children’s Hospital has many resources on cancer treatment for children and families.1  The UNC Lineberger Comprehensive Cancer Center provides information to families about OS with links to other organizations that support patients with sarcomas.19  The Ronald McDonald House of Chapel Hill is another place for families to seek support and comfort.20

 A Childhood Cancer Survivor Study of 528 patients with bone tumors reported overall excellent functioning and quality of life.23  Promoting physical activity and fitness can play a role in preserving this outcome. To support continued activity, fitness and wellness to adolescents in their recovery from OS several resources might be explored. Children who have undergone limb sparing surgery exhibit decreased physical activity compared to typically developing age matched peers.21  Beads of Courage is one program where children can collect beads for procedures and interventions they experience while in the hospital.22 Earning beads for exercising is motivating for some children.22

Camp Carefree is a summer camp which has a week dedicated to cancer survivors. This would be an appropriate resource for children with OS to be active and seek support from other children who have survived cancer. Bridge II Sports is another local organization that could provide sports and recreational activities to adolescents recovering from OS.24

For children with lower extremity amputations or limb salvage, there are many opportunities to participate in local biking, walking and running events. Pedal for Pediatrics and The Get Heeled 5K actually raise funds for pediatric cancer patients and their families at UNC.25 Participation in these local resources may facilitate ongoing activity and participation in adolescents with OS.

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