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.

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.

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).

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.

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.)
Etiology

The cause of ESFT is unknown. It does not appear to be inherited (Grier, 1997). Radiation exposure and other environmental factors do not appear to be associated with the disease (Grier). The family of tumors is not commonly associated with congenital diseases (Pizzo & Poplack, 2005). The tumors may occur as secondary malignancies but the incidence is low.

Clinical Presentation

The most common presenting symptoms of ESFT are pain and swelling. Local pain is seen in 85% of children, and local swelling is seen in 60% (Lanzkowsky, 2005). Other common presenting symptoms include a palpable mass and pathologic fracture (see Figure 1). Systemic signs and symptoms such as fever, weight loss, and an increased sedimentation rate (in as many as 50% of cases) also may be seen upon presentation (Pizzo & Poplack, 2005). The systemic symptoms often lead to differential diagnosis, including infection, namely osteomyelitis. Ewing’s sarcoma can originate in any bone in the body or in extraosseous sites (soft-tissue tumors) (Horowitz et al., 1992). Common primary sites of origin are listed in Figure 2. Patients also can present with symptoms from a metastatic site rather than the primary site. For example, back pain upon presentation may signify spinal cord compression secondary to the primary tumor or a metastatic spinal tumor (Pizzo & Poplack).

Also common is a delay between the onset of symptoms and diagnosis (Pizzo & Poplack, 2005). This may be caused by intermittent pain at the site of the tumor, or by a mass that is not palpable until it is fairly large in some locations. Forty-eight percent of 331 subjects treated on the Intergroup Ewing’s Sarcoma Study 1 showed a three-month delay between presentation of symptoms and diagnosis (Pizzo & Poplack).

Common sites of metastatic spread of ESFT are lung, bone, and bone marrow, with approximately 25% of patients presenting with metastatic disease at time of diagnosis. To rule out metastasis, healthcare professionals should obtain a bone scan, computed tomography scan of the chest, and bilateral bone marrow biopsies and aspirates prior to the start of treatment. The site of the primary tumor shows a relationship to the presence of metastatic disease at the time of diagnosis (Lanzkowsky, 2005) (see Table 1).

Pathology

Ewing’s sarcoma is most often a tumor of bone (osseous) but can occur in soft tissue (extraosseous). ESFT is referred to as small round-cell tumors. Other types of cancer included in the group are neuroblastoma, rhabdomyosarcoma, and non-Hodgkin lymphoma. Differentiating Ewing’s sarcoma from other small, round-cell tumors sometimes is difficult, especially with the soft-tissue type of Ewing’s sarcoma (Grier, 1997). ESFT is thought to be of parasympathetic nerve origin. The classic Ewing’s sarcoma tumor consists of undifferentiated cells that often have scant cytoplasm, are round or oval, have hyperchromatic nuclei, and test positive for glycogen (Pizzo & Poplack, 2005).

PNET, on the other hand, shows apparent differentiation. Both Ewing’s sarcoma and PNET are thought to arise from neoplastic growth of the same precursor neural cell in either bone or soft tissue (Horowitz et al., 1992). The histologic difference between the two types of tumor is seen in their degree of differentiation. Whether a difference exists in outcome between standard Ewing’s sarcoma histology and neuroectodermal differentiation remains controversial (Grier, 1997).

Cytogenetic testing reveals a characteristic translocation, t(11; 22) in 88%–95% of tumors in ESFT (Pizzo & Poplack, 2005).
The treatment for ESFT consists of a multimodal approach: chemotherapy, radiation therapy, and surgery. Systemic chemotherapy is for eradication of microscopic disease, and radiation therapy and surgery are for control of the primary lesion. The ultimate goal of treatment is cure while preserving function and reducing late effects.

Historically, local control (radiation therapy and surgery) was the mainstay of therapy, but more than 90% of patients eventually died from metastatic disease (Pizzo & Poplack, 2005). Healthcare professionals assumed that every patient had microscopic disease at diagnosis (Pizzo & Poplack), so systemic chemotherapy was added to local therapy (Intergroup Ewing’s Sarcoma Study One) beginning in 1973; this not only improved overall survival rates but also showed improvement in local control of the primary tumor after radiation therapy (Grier, 1997) (see Table 2). The trial included a randomization of three groups: (1) vincristine, actinomycin-D, and cyclophosphamide alone; (2) vincristine, actinomycin-D, cyclophosphamide, and doxorubicin; and (3) vincristine, actinomycin-D, and cyclophosphamide with bilateral pulmonary irradiation (Nesbit et al., 1990). Of the three groups, vincristine (2 mg/m²), actinomycin-D (2 mg/m²), cyclophosphamide (1.200 mg/m²), and doxorubicin (75 mg/m²) was superior, with a five-year event-free survival of 60%. Beginning in the 1980s, ifosfamide (1,800 mg/m²) and etoposide (100 mg/m²) were used for patients with relapsed ESFT. The studies then led to the collaborative children’s oncology groups initiating randomized, controlled trials.

The first National Cancer Institute protocol (INT-0091) opened in 1988 and closed to accrual in 1992; 530 patients were enrolled, 518 of whom met eligibility criteria, including being 30 or younger. Of the 518, 120 had metastatic disease and 398 had non-metastatic disease (Grier et al., 2003). Of the metastatic group, 62 were randomly assigned to the standard treatment arm and 58 to the experimental treatment arm. Of the non-metastatic group, 198 were randomly assigned to the experimental treatment arm and 200 to the standard arm. The protocol’s two arms were (1) vincristine, actinomycin-D, cyclophosphamide, and doxorubicin; and (2) vincristine, actinomycin-D, cyclophosphamide, and doxorubicin alternating with courses of ifosfamide and etoposide. The schema of the protocol involved 17 courses occurring every three weeks for a total duration of 49 weeks, with the local control occurring at week 12 (i.e., radiation and/or surgery). Five-year event-free survival for patients in the experimental group was superior to that of the standard therapy group, 69% versus 54%. (Grier et al.). The regimen including alternating courses of ifosfamide and etoposide was

### Prognosis

The prognosis of ESFT varies with site of the primary tumor, presence of metastases, and tumor size (see Figure 3 for prognostic factors). The overall five-year disease-free survival rate for localized Ewing’s sarcoma treated with surgery, radiation, and multiagent chemotherapy is 65%–76% (Lanzkowsky, 2005). With metastases present at diagnosis, the five-year disease-free survival rate drops to 30%; limited improvement of survival rates has been made for metastatic Ewing’s sarcoma (Miser et. al., 2004). Those with gross soft-tissue extension of the primary tumor have an increased incidence of local recurrence and subsequent metastases as well as a decreased five-year disease-free survival rate. Tumors that extend into soft tissue also have an increased incidence of metastases at diagnosis (Lanzkowsky). Little has been published about the outcome of adults who have ESFT (Martin & Brennan, 2003).
superior to vincristine, actinomycin-D, cyclophosphamide, and doxorubicin alone. The addition of ifosfamide and etoposide to the previous standard regimen significantly improved the outcome for patients with non-metastatic ESFT (Grier et al.).

The next step to improve survival rates involved research into the drugs and the times when they are given. Because ESFT is sensitive to alkylating agents that have a step-dose response curve, dose intensification is an alternative (Marina et al., 1999). This is considered interval compression, which, in theory, is increasing the total dose of drugs administered over shorter periods of time, which should result in greater cell kill, thus a better cure rate. From that conclusion, the collaborative children’s oncology groups developed protocols that compared treatment given in 30 weeks as compared to the standard schema of 48 weeks. In that study, the type of local control also was compared with the groups, including radiation, subtotal resection with radiation, and total surgical resection. The theory also involved limiting time in which partially drug-resistant cells have to recover (Womer, Daller, Fenton, & Miser, 2000).

Local Control

The use of highly sophisticated diagnostic methods (including computed tomography, three-dimensional computed tomography, magnetic resonance imaging and three-dimensional magnetic resonance imaging) has led to improved preoperative planning for local control (Sluga et al., 2001). Also, the extensive development of protheses and intraoperative handling of Ewing’s tumors have improved overall survival and disease-free survival (Sluga et al.). Location of the primary tumor continues to be the most important factor when determining prognosis; nonpelvic tumors have better outcomes (Sluga et al.). The treatment options for local control continue to be radiation or surgery or both. The most influential factor related to surgical success is the resection margins; the wider the resection margins, the lower the rate of local recurrences (Sluga et al.).

The expertise in surgical options also helps to determine histologic response, further use of systemic chemotherapy, and overall survival rate. Surgical options include amputation and limb salvage with or without rotationplasty. Prostheses for children have been developed, which has aided in surgical advancements (Sluga et al., 2001).

In surgery, the goals include clear and wide margins, indicating healthy tissue, and maintaining optimal function. All of this is dependent on the tumor size, location, and extent of disease outside the primary site.

Surgery also comes with its host of complications, including postoperative pain, infection, incision and wound healing problems, and mobility changes. Surgery also can cause other complications, such as phantom pain in those who have amputations, when the body believes or perceives that the body part is still there. Interventions including pharmacologic (e.g., gabapentin) and nonpharmacologic (e.g., guided imagery) techniques are very specific to managing phantom pain. For patients undergoing limb salvage, a potential complication is “nonunion of the bones,” when the bone does not heal, increasing the risk of fractures and prohibiting the growth of healthy tissue (Baggott, Kelly, Fochtman, & Foley, 2002).

Radiation

The choice of local control is based on the ability to achieve clear margins and balance functional and cosmetic surgery versus radiation and its side effects (Gibbs, Tuamokumo, & Yock, 2006). Before radiation, a prechemotherapy magnetic resonance imaging scan is performed to determine tumor extent and treatment volume. The recommended radiation doses are 55.8 Gy for gross residual disease and 50.4 Gy for microscopic disease after surgical resection. The amount of normal tissue that is affected will help determine functional outcomes after radiation.

The side effects and complications that can occur include skin irritation, postsurgical wound complications, limb-length discrepancies, soft tissue and muscle changes, and secondary malignancies (dose related). Radiation continues to remain a vital part of treatment for ESFT.

High-Dose Chemotherapy With Stem-Cell Rescue

The prognosis for children and young adults with Ewing’s sarcoma steadily improved after the introduction of multimodality therapy in the 1960s and 1970s. However, in 1980, a group of patients at high risk for treatment failure could be identified. The

Table 2. Historical Perspective of Ewing’s Sarcoma Treatment and Survival

<table>
<thead>
<tr>
<th>TIME PERIOD</th>
<th>TREATMENT</th>
<th>SURVIVAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>1953</td>
<td>Local control (surgery or radiation)</td>
<td>10%</td>
</tr>
<tr>
<td>1973</td>
<td>Systemic chemotherapy added to local control (standard regimen: vincristine, Adriamycin, and cyclophosphamide)</td>
<td>50%–60% (five-year event-free survival)</td>
</tr>
<tr>
<td>1980–1990s</td>
<td>Addition of systemic chemotherapy to standard regimen (ifosfamide and etoposide)</td>
<td>69%</td>
</tr>
<tr>
<td>2000–2005</td>
<td>Concept of interval compression added to treatment regimens</td>
<td>&gt; 70% and increasing</td>
</tr>
</tbody>
</table>

Note. Based on information from Grier et al., 2003; Pizzo & Poplack, 2005.

Figure 3. Prognostic Factors of Ewing’s Sarcoma Family of Tumors

Note. Based on information from Gibbs et al., 2006.
Clinical Journal of Oncology Nursing  •  Volume 12, Number 1  •  Ewing's Sarcoma Family of Tumors 93

most important adverse prognostic factor is clinically detectable metastatic disease, which is present in 20%–30% of patients at diagnosis. As many as one-third of patients whose metastases are limited to the lungs or pleura become long-term survivors with conventional multimodal therapy. Patients with bone or bone marrow metastases have an even worse prognosis (Pizzo & Poplack, 2005). Recognition of this problem, combined with improvements in supportive care, has led to dose intensification as high-dose chemotherapy with stem-cell rescue in this population of patients. Literature is limited regarding high-dose chemotherapy with stem-cell rescue in high-risk patients with Ewing’s sarcoma. A few reports are available on the use of high-dose therapy with stem-cell rescue in patients with metastatic bone or bone marrow involvement. In a review of the literature and a report from Memorial Sloan-Kettering Cancer Center, Kushner and Meyers (2001) reported a cohort of 21 patients with newly diagnosed Ewing’s sarcoma metastatic to the bone marrow or bone who underwent high-dose chemotherapy with stem-cell rescue. All patients received induction therapy with cycles consisting of cyclophosphamide (4.2 g/m²), doxorubicin (75 mg/m²), and vincristine (2 mg/m²) alternating with cycles of ifosfamide (9 g/m²) and etoposide (500 mg/m²). Patients who achieved complete remission or very good partial remission proceeded to consolidation with high-dose chemotherapy and stem-cell rescue. Two ablative regimens were used: total body irradiation (15 Gy hyperfractionated) and melphalan (180 mg/m²) or thiotaopo (900 mg/m²) and carboplatin (1,500 mg/m²). Eight patients received the total body irradiation and melphalan regimen; four relapsed 27 months after transplantation; two patients died early from toxicity; one died from pulmonary failure 17 months after transplantation with no evidence of cancer. One remaining patient remains in complete remission more than seven years after transplantation. The three patients treated on the carboplatin and thiotaopo regimen relapsed three to four months after transplantation.

In reviewing the literature, patients with metastatic Ewing’s sarcoma to the bone or bone marrow show similar poor results. The results of the literature search and the Memorial Sloan-Kettering experience suggest that relapse remains the main obstacle to success in this high-risk group of patients. This select group of patients awaits the introduction of novel therapies to have an impact on prognosis and long-term survival (Kushner & Meyers, 2001).

Nursing Care of Patients With Ewing’s Sarcoma Family of Tumors

Children and young adults with Ewing’s sarcoma face many physical and psychological challenges, starting with their illness through treatment and in the future as survivors of childhood cancer. Because of the aggressive multimodal nature of treatment for Ewing’s sarcoma, children and young adults experience significant toxicities.

Bone Marrow Suppression

Bone marrow suppression is one of the most common dose-limiting toxicities of chemotherapy, including anemia, neutropenia, and thrombocytopenia. Bone marrow suppression may occur as a result of disseminated disease to the bone marrow or, in most cases, because of the aggressive treatment course. Neutropenia is a decrease in the number of neutrophils that fight infection and is the most severe consequence of bone marrow suppression. Children and young adults with severe neutropenia (absolute neutrophil count < 500) are at risk for life-threatening infections, whether bacterial, fungal, or viral in etiology. Any patient with Ewing’s sarcoma who presents with fever or other signs of infection warrants a thorough physical assessment, an essential nursing role. The evaluation includes, but is not limited to, hemodynamic instability monitoring, peripheral pulses, skin color, perfusion, and vital signs; skin assessment for abnormal integrity, erythema, swelling or drainage, and micturition; visual inspection of the central venous catheter site; assessment for any respiratory compromise such as presence of a cough and lung auscultation; examination of the ears, nose, throat, and mouth for rhinorrhea, tenderness, erythema, or stomatitis; abdomen auscultation and assessment of the abdomen for tenderness; and visual inspection of the rectum for perirectal changes. Nurses also must communicate pertinent laboratory values and physical findings to the treatment team and recognize the subtle signs of infection and evidence of impending septic shock.

During preparation for discharge from routine chemotherapy admissions, homecare teaching and instructions must communicate clearly that fever in this population is a medical emergency. Parents should be instructed to call if a patient experiences a temperature higher than 38°C (100.4°F) on two occasions in 24 hours or a one-time temperature equal to or greater than 38.5°C (101.3°F) in a 24-hour period. Families also must be instructed to report shaking chills, signs of respiratory compromise, changes in a child’s level of consciousness, or any general change in a child or young adult’s condition, even if not associated with a fever. The keys to the management of febrile neutropenic patients are early assessment and intervention.

Children and young adults with anemia present with decreased energy, fatigue, pallor, tachypnea, tachycardia, and/or headache. The standard transfusion volume is 10–20 ml/kg of packed red blood cells. The packed red blood cells should be cytomegalovirus negative, irradiated, and, if needed, leukoreduced (patient specific). The hemoglobin values used to determine criteria for transfusion vary by institution. Families should be taught to recognize the signs and symptoms of anemia and report their findings to the treatment team.

Thrombocytopenia is defined as having fewer than 100,000/mm³ circulating platelets. It can be caused by malignant cells in the marrow, myelosuppressive therapy, or increased platelet consumption, which can happen in situations such as disseminated intravascular coagulation or fever (Baggott et al., 2002). Spontaneous bleeding generally is associated with a platelet count of < 10,000/mm³. Children may have presenting symptoms such as petechiae, increase bruising, epistaxis, and mucosal bleeding (Baggott et al.). Assess the child’s mouth, skin, nose, sclera, and output (i.e., urine, stool, and emesis) for signs of bleeding or use appropriate testing strategies for microscopic bleeding. Clearly, transfusions are indicated when a child is bleeding and may be given prophylactically when a child’s platelet count is 10,000–20,000 cells/mm³. Controversy remains regarding the transfusion of platelets and whether they should be given prophylactically because of the risk of alloimmunization, transfusion reactions, and hepatitis (Pizzo & Poplack,
Stomatitis

Inflammation of the mucous membranes, somatitis, is a common dose-limiting toxicity induced by chemotherapy, radiotherapy, and neutropenia. Cell destruction by chemotherapy or radiation and inadequate production of new cells result in a loss of mucosal integrity (Baggott et al., 2002). The consequences of somatitis include painful lesions that often cause decreased oral intake and nutrition. Patients often require parenteral nutrition for pain management. The compromised oral mucosa provide a portal of entry for microorganisms, which, when accompanied by concomitant neutropenia, can result in significant infections. Recovery from somatitis often occurs as neutropenia resolves. The onset, duration, and severity of somatitis are related to the agent, dose, and duration of chemotherapy administration. Stomatitis can range from mild erythema of the mucosal membranes to ulcerations; signs and symptoms include pain (mouth and rectal), difficulty swallowing, withholding of stool, thick oral secretions, white patches, cracked or dried lips, and drooling. Fungal and viral infections of the mucous membranes can occur, especially in the mouth and esophagus. The most common fungal infection is caused by Candida albicans and presents as white plaques with indurated borders. The most common viral infection is herpes simplex virus, which presents as painful blisters on the lips or anywhere in the mouth, usually accompanied by a yellowish brown membrane.

The current treatment for chemotherapy-induced oral somatitis continues to focus on symptoms with mouth care, analgesics, and nutritional support. Recommendations include oral care protocols with education, using an objective oral assessment tool, and the inclusion of dental professionals. The consensus is that routine, basic oral care is required for the prevention and management of oral somatitis. An example of a good oral hygiene program used in many pediatric practices includes flossing daily (unless contraindicated by a physician), brushing morning and night and after meals, cleansing the oral cavity with an appropriate cleansing agent before bed and after meals, and avoid irritating agents such as alcohol, astringents, and spicy foods. The recommendation is for a bland rinse to be used, which will help remove debris and aid with oral hydration (Harris, Eiler, Cashavelly, Maxwell, & Harriman, 2006). Maintaining adequate nutrition and hydration is a challenge for children and young adults with moderate to severe somatitis. A soft diet with cool and bland foods may be the most widely accepted form of nutrition. Ice chips and ice pops can be soothing and helpful in hydrating and maintaining moisture on the lips and mucous membranes. Patients’ nutritional status must be monitored closely. Recording intake, output, and weight is necessary. Education regarding oral hygiene begins at the time of diagnosis and includes the expected time of onset for somatitis, the signs and symptoms, methods of good oral hygiene, nutrition, and potential complications such as infection, bleeding, and dehydration (Baggott et al., 2002).

Gastrointestinal Toxicities

Nausea and vomiting are among the most common side effects of cancer treatment and may be acute, delayed, or anticipatory. A variety of techniques may be used to control or prevent nausea and vomiting; the most common is the administration of antiemetics. Numerous categories of antiemetics are used in children and young adults with cancer: serotonin antagonists (ondansetron, granisetron, and dolasetron), phenothiazines (chlorpromazine and prochlorperazine), corticosteroids (dexamethasone and methylprednisolone), benzodiazepine (lorazepam), antihistamines (diphenhydramine and hydroxyzine), procainamide derivatives (metoclopramide), and cannabinoid tetrahydrocannabinol. Nonpharmacologic techniques can be combined with antiemetic therapy depending on patient age and preferences; they include but are not limited to guided imagery, music therapy, massage therapy, exercise, and ginger (Tipton et al., 2005).

Diarrhea is defined as an abnormal increase in the quantity, frequency, and liquidity of stool (Baggott et al., 2002). Diarrhea can lead to severe dehydration, renal insufficiency, electrolyte imbalances, and impaired skin integrity. The clinical presentation of diarrhea illness depends largely on the type and cause. The goal of diarrhea management is to restore normal bowel habits, maintain adequate nutrition, restore fluid electrolyte balance, and protect skin integrity. The management of diarrhea is directly related to its cause; therefore, establishing the underlying cause is the first step in effective treatment (Baggott et al.). Nurses must maintain strict intake and output measurements, obtain patient weight daily or more frequently as indicated, and provide skin care as needed.

Constipation is a decrease in the normal frequency of stool production and may include a decline in the normal production of stool, change in the characteristics of stool, hypoactive bowel sounds, distended or firm abdomen, diffuse abdominal tenderness, decreased appetite, straining with attempted bowel movements, and pain associated with defecation. With early assessment and intervention, constipation in patients with cancer often is manageable and preventable (Baggott et al., 2002). Obtaining a baseline assessment of a patient’s normal bowel habits is important. Accurate records should be maintained, including the frequency, consistency, and amount of stool output. A thorough abdominal assessment includes bowel sounds, the degree of abdominal distention, and the presence of pain or cramping. Because constipation also can be associated with straining and passing hard stools, the perirectal area must be inspected thoroughly for the presence of hemorrhoids, skin breakdown, rectal fissures, or bleeding. Nurses must advocate for prophylactic measures to prevent constipation and have an important role in the prevention, diagnosis, and treatment of constipation.

Nutritional support is an integral component of care that can directly affect clinical outcomes. Malnutrition is reported to occur in 8%–32% of the children treated for cancer (Tyc, Valleurunga, Mahoney, Smith, & Mulhern, 1995). Mechanical barriers, systemic and local effects of disease process, side effects of treatment, psychosocial reactions to the disease process, or a combination of these factors all contribute to alterations in nutrition. Mechanical barriers may interfere with oral intake.
by limiting the ability to eat or chew. Anorexia and cachexia are among the most common nutritional complications of cancer and its treatment (Tisdale, 1991). Factors contributing to anorexia may include nausea, vomiting, taste diversion, early satiety, anxiety, depression, and environmental changes (Boggott et al., 2002). Interventions for anorexia involve counseling patients with individualized diets designed to promote nutritional restoration and prevent further nutritional deficits. “Cancer cachexia is a syndrome that develops secondary to the progressive growth of malignant process and limits the quality of life, length of survival, and treatment options. The clinical features include muscle wasting, anorexia, weakness, anemia, and metabolic deficit” (Boggott et al., p. 297). Goals of treatment include restoring and promoting growth and development and minimizing side effects. Nutritional screening must take into account a patient’s weight, food intake, treatments that affect nutrition, functional status, physical examination, and biochemical indicators of protein scores (albumin and prealbumin levels) (Pizzo & Poplack, 2005). To prevent malnutrition and subsequent treatment complications, nutritional interventions should be initiated early and may include diet modifications, oral supplements, enteral feedings, parenteral nutrition, and medications to stimulate appetite (megestrol). Controversy often exists over the choice of enteral versus parenteral nutrition. Unless contraindicated, the enteral approach to nutritional support is more advantageous because continued stimulation of the gut prevents atrophy of the villi of the intestinal wall and minimizes changes in bacterial flora and the risk for sepsis (Bloch, 2000).

### Table 3. Late Effects of Treatment by System

<table>
<thead>
<tr>
<th>SYSTEM</th>
<th>CAUSATIVE AGENTS</th>
<th>POTENTIAL EFFECTS</th>
<th>INTERVENTIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Genitourinary</td>
<td>Cyclophosphamide</td>
<td>Hemorrhagic cystitis</td>
<td>Education about reporting hematuria</td>
</tr>
<tr>
<td></td>
<td>Ifosfamide</td>
<td>Bladder fibrosis</td>
<td>Maintain adequate hydration.</td>
</tr>
<tr>
<td></td>
<td>Radiation (to affected area)</td>
<td>Kidney dysfunction</td>
<td>Monitor blood pressures.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Referral to urologist</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Anthracyclines</td>
<td>Cardiomyopathy</td>
<td>Routine tests: echocardiogram and multiple gated acquisition scan (total dose dependent)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Exercise intolerance</td>
<td>Education: dietary management</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hypertension</td>
<td>Exercise program</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Avoid alcohol and tobacco.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Avoid heavy lifting.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Electrolyte supplementation</td>
</tr>
<tr>
<td>Reproductive and endocrine</td>
<td>Cyclophosphamide</td>
<td>Sexual dysfunction</td>
<td>Routine laboratory values: follicle stimulating hormone, luteinizing hormone, spernum analysis, triiodothyronine, thyrline, and thyroid stimulating hormone</td>
</tr>
<tr>
<td></td>
<td>Ifosfamide</td>
<td>Growth abnormalities</td>
<td>Sperm donation (men) (before chemotherapy and radiaion if area is included in field)</td>
</tr>
<tr>
<td></td>
<td>Radiation</td>
<td>Sterilization</td>
<td>Education during treatment</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Menstrual cycle changes</td>
<td>Hormonal support</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Referral to endocrinologist or fertility specialist</td>
</tr>
<tr>
<td>Hematopoietic</td>
<td>Anthracyclines</td>
<td>Acute myeloid leukemia</td>
<td>Routine physical examination</td>
</tr>
<tr>
<td></td>
<td>Epipodophyllotoxins (etoposide)</td>
<td></td>
<td>Routine complete blood count with differential</td>
</tr>
<tr>
<td></td>
<td>Radiation to marrow-containing bones</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Central nervous</td>
<td>Radiation to spine</td>
<td>Paresthesias</td>
<td>Computed tomography and magnetic resonance imaging</td>
</tr>
<tr>
<td></td>
<td>Surgical manipulation near spine</td>
<td>Chronic pain</td>
<td>Referral to neurology</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Radiation to abdomen</td>
<td>Fibrosis and strictures</td>
<td>Growth checks</td>
</tr>
<tr>
<td></td>
<td>Concomitant use of actinomycin and doxorubicin</td>
<td>Malabsorption</td>
<td>Routine laboratory values (chemistries and complete blood count)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Anorexia, nausea, vomiting</td>
<td>Referral to gastroenterologist</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Change in bowel habits</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Food intolerance</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Jaundice</td>
<td></td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>Radiation to long bones, spine, any growing bone/muscle area</td>
<td>Muscle or bone asymmetry</td>
<td>Bone age films</td>
</tr>
<tr>
<td></td>
<td>Amputation or limb salvage</td>
<td>Muscle or bone hypoplasia</td>
<td>Standing and sitting heights</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Limb length discrepancy</td>
<td>Encourage physical activity.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Alterations in growth</td>
<td>Encourage weight control.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Gait changes</td>
<td>Referral to orthopedist</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Functional deficits</td>
<td>Referral to physical and occupational therapy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Chronic pain</td>
<td></td>
</tr>
</tbody>
</table>
Fatigue is a major complication of cancer treatment. Causes include treatments for malignancy; the side effects of low blood counts, fever, and pain; and too many activities (Hockenberry-Eaton et al., 1998). A major cause of fatigue in the hospital is when hospital staff, medical equipment, and other noises interrupt patients’ sleep. Worries and fears also may cause fatigue. A variety of nursing interventions can be used to help alleviate fatigue in children and young adults who are being treated for Ewing’s sarcoma. The Oncology Nursing Society’s Putting Evidence Into Practice® document on fatigue (Mitchell, Beck, Hood, Moore, & Tanner, 2005) recommends the following interventions: prescreening for contributing factors, energy conservation, exercise and activity management, optimizing sleep quantity and quality, relaxation, and massage therapy. Numerous other optional interventions exist, but those listed here are supported by evidence and systematic review. For children and young adults, healthcare professionals should consider respecting sleep time and decreasing interruptions, noise, and waiting times.

A well-known side effect of chemotherapy is alopecia; it occurs inevitably and usually over several treatment courses. Prepare adolescents and young adults and assist them with the loss. It can be quite devastating to female adolescents, as egocentrism and identity are a large part of their development during this time.

Cardiovascular Toxicities

Cardiac complications related to therapy for ESFT may be categorized as acute or chronic and may arise depending on the type of chemotherapy received, whether radiation was received in the mediastinal area, or whether disease exists within the heart itself (Baggott et al., 2002). One of the worrisome cardiac complications that can affect patients with ESFT is cardiomyopathy from the anthracycline class of chemotherapeutic agents. Patients receiving anthracyclines have a lifelong cumulative dose (450 mg/m² for doxorubicin and daunorubicin and 125 mg/m² for idarubicin) (Albin, 1997). Signs and symptoms include those of congestive heart failure, such as shortness of breath, cough, weight gain or edema, and activity intolerance. Patients receiving anthracycline therapy must have routine cardiac studies before, during, and after therapy, most of which are outlined in the specific treatment protocol.

Neurotoxicities

One of the major neurotoxicities that can result from treatment for ESFT is peripheral neuropathy from vinca alkaloids, primarily vincristine. Peripheral neuropathy can present as numbness or tingling in the limbs or on examination as depressed or absent deep tendon reflexes (Baggott et al., 2002). Patients may report difficulty walking or new onset of falling. This can lead to foot drop or parathesias; therefore, prompt identification is important to begin appropriate treatment (Visovsky, Collins, Hart, Abbott, & Aschenbrenner, 2006).

Late Effects of Therapy and Psychological Implications

With the increasing progress of treatment of childhood cancer and increasing number of survivors of childhood cancer, the long-term (direct and indirect) sequelae of therapy must be addressed. Beyond monitoring for recurrence, caring for childhood cancer survivors must include evaluation of potential long-term effects, health education, and promotion of health screening (Nagarajan et al., 2003). Late effects for patients with ESFT are attributed to all modalities of therapy (Bottomley & Kassner, 2003) (see Table 3). Survivors of ESFT can suffer from multiple psychological sequelae, including but not limited to fear of recurrence, sense of physical damage (related to local therapy of limb salvage or amputation), post-traumatic stress disorder, anxiety, depression, difficulties with interpersonal relationships, and financial and employment issues. Survivors of ESFT have more difficulty adapting to adulthood; therefore, healthcare professionals must provide appropriate supportive services and counseling for those in need of psychosocial interventions (Nagarajan et al.).

Nurses must be able to identify patients at risk for late effects from the sequelae of their treatments. Nurses should offer resources and education related to patients’ disease history, treatments, and possible health deficits. These are new challenges to nurses, because ESFT once was a noncurable disease.

Author Contact: Meredith Lahl, RN, MSN, CNS, CPON®, can be reached at lahlm@ccf.org, with copy to editor at CJONEditor@ons.org.

References


