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Hematologic Childhood Cancers: An Evidence-Based Review

Treatments, supportive measures, and essential patient teaching points.

ABSTRACT: Every year in the United States, thousands of children and adolescents are diagnosed with a hematologic cancer. That diagnosis and the prescribed course of treatment profoundly affect both the child and the family. This article provides a brief overview of the therapies used to treat such cancers, describes the presentations and diagnoses of the various hematologic cancers, and explains the treatments specific to each. Nursing care of the child and family is discussed, with an emphasis on education and supportive care.

Keywords: acute lymphoblastic leukemia, acute myelogenous leukemia, hematologic cancer, Hodgkin lymphoma, leukemia, lymphoma, non-Hodgkin lymphoma, oncology, patient education, pediatric cancer

Childhood cancer rates have been on the rise for several decades.[1] According to the Centers for Disease Control and Prevention (CDC) WONDER online database, in 2014 (the most recent year for which data are available), 15,190 new cases of cancer were diagnosed in U.S. children and adolescents under the age of 20, 41.5% (6,309) of which were leukemias or lymphomas.[2] Clinical trials sponsored by centers affiliated with the Children's Oncology Group of the National Cancer Institute (NCI), which treat more than 90% of childhood cancers in the United States, as well as the collaborative international efforts of other cancer and pediatric health centers, have significantly improved childhood cancer therapy and outcomes.[3,4] Over the past 45 years, five-year survival rates among U.S. children with cancer have risen from 58% to more than 80%,[1] although cancer continues to be the leading cause of childhood death by disease beyond infancy.[5] Based on post-2009 childhood cancer incidence, prevalence, and survival rates, the number of U.S. childhood cancer survivors is expected to approach 500,000 by 2020.[6] Late effects of childhood cancer and late complications of the therapies used to treat it are common. Between 60% and 90% of childhood cancer survivors develop one or more chronic health conditions during adulthood, while 20% to 80% experience severe or life-threatening complications as adults.[7]

It is important for all nurses to have a basic knowledge of childhood cancers, because in any work setting a nurse may care for a child experiencing complications of cancer or its treatment or an adult survivor of childhood cancer experiencing late effects. This article focuses specifically on hematologic cancers, which are among the most common types of childhood cancer (see Table 1[8-13]). It discusses the approaches used to treat such cancers, explains which treatments are indicated for the various types of leukemia and lymphoma, and describes the education and supportive care both the child and the family require.

By Jessica Lynne Spruit, DNP, RN, CPNP-AC

Photo courtesy of the Children's Hospital of Michigan.

APPROACHES USED TO TREAT HEMATOLOGIC CANCERS IN CHILDREN

Hematologic cancers in children are now treated with a variety of modalities that have been developed or fine-tuned in response to a continually expanding knowledge base in the fields of genetics and cancer biology. These treatment approaches, which have reduced both treatment-related toxicities and late effects of cancer treatment, include multiagent chemotherapy, radiation therapy, targeted therapies, and hematopoietic stem cell transplantation (HSCT).

Multiagent, or combination, chemotherapy uses antineoplastic agents with dissimilar toxicity profiles and different mechanisms of action to reduce adverse effects while improving the likelihood of treatment effectiveness. Depending on the specific agents used, as well as such variables as patient organ function and body weight, and any other therapies taken, administration may vary. Chemotherapy may be administered by mouth, intermittent or continuous infusion lasting up to several days, or intramuscular or subcutaneous injection. Nurses who administer chemotherapeutic agents to children must be aware of potential adverse effects and the medication-specific patient monitoring that is required during administration.

Radiation therapy may include the administration of photons (high energy X-rays that can damage DNA in targeted tissue), electrons (negatively charged particles that are absorbed in tissue at a limited depth), or protons (high-energy particles associated with reduced damage to surrounding tissues owing to decreased distribution on entry and exit).[14] Radiation therapy may be used in conjunction with chemotherapy in children with hematologic cancers, despite the risk it poses to healthy tissues exposed during treatment. But advances in chemotherapy have allowed practitioners to reduce radiation dosage for patients who require it.[15]

Targeted therapies are some of the most exciting and promising treatments in childhood cancer. Unlike traditional chemotherapy drugs, which kill all rapidly dividing cells, targeted cancer therapies are able to target malignant cells by seeking out genetic markers that differentiate them from normal cells.[16] Targeted therapies are divided into two classes: drugs and biologics. Biologic therapies include monoclonal antibodies, which are synthetic antibodies that work in a variety of ways. Some

block antigens on cancer cells that allow tumors to proliferate; others attach to antigens or proteins on cancer cells, enabling the patient's immune cells or a chemotherapy medication to target tumors.[17] Biologics also include adoptive cellular therapies, which use modified T cells to help the patient's immune system fight disease, such as T cells altered to express chimeric antigen receptors, which have been created in a laboratory to bind to proteins found on cancer cells. Clinical trials of such targeted therapies are ongoing and have the potential to reduce adverse effects and long-term complications in children and adolescents with hematologic cancers.

HSCT. Two types of HSCT are used to treat childhood blood cancers: autologous, which uses the patient's own stem cells, and allogeneic, which uses stem cells from a healthy donor.

Autologous stem cells are harvested from the patient between cycles of chemotherapy and reinfused after a regimen of high-dose chemotherapy (often called the "preparative" or "conditioning" regimen).[18] Autologous HSCT allows patients to receive doses of chemotherapy that may otherwise lead to bone marrow failure, and is considered a "stem cell rescue" after such high-dose therapy.[19]

Allogeneic stem cells are collected from healthy related or unrelated donors. Sources may include peripheral blood, bone marrow, or blood collected from the umbilical cord and placenta (after the umbilical cord is clamped). They are infused after a preparative regimen of chemotherapy and immunosuppression.

Because there are several complications specific to HSCT, including graft-versus-host disease and infection related to immunosuppression, it is considered a high-risk treatment and is reserved for advanced or recurrent disease.

PRETREATMENT CONCERNS

Before initiating therapy for childhood cancer, health care providers must consider the patient's organ function and ability to tolerate planned therapies. The patient's treatment plan determines necessary pretreatment testing, which should be specified in either the treatment protocol or the institution's standard monitoring recommendations. Such testing may include an echocardiogram to gauge cardiac function, an audiogram to appraise hearing, or calculation of glomerular filtration rate to evaluate kidney function.

Fertility preservation is another important pretreatment consideration. If initiation of chemotherapy is not emergent, nurses should discuss the possibility of sperm cryopreservation for male patients and oocyte or embryo cryopreservation for female patients. Additional experimental procedures are also available at select centers.[20]

CHILDHOOD LEUKEMIA

Leukemia is the most common type of cancer diagnosed in U.S. children and adolescents under age 20. White blood cells, specifically lymphocytes, rapidly proliferate in dysfunctional bone marrow. The two most common types of leukemia in children are acute lymphoblastic leukemia (ALL) and acute myelogenous leukemia (AML). Additional specified and unspecified types of leukemia occur in children, but the incidence of these subtypes is much lower.[2]

Table 1. The Most Common Childhood Hematologic Cancers[8-13]

Type of Cancer	Gender Distribution	Peak Incidence (by age)	Diagnostic Evaluation
Acute lymphoblastic leukemia	Boys > girls	1–4 years	Bone marrow morphology to determine percentage of blasts; > 25% is diagnostic.
Acute myelogenous leukemia	Boys = girls	First 12 months	Bone marrow morphology to determine percentage of blasts; ≥ 20% is diagnostic. If the procedure is contraindicated, initial diagnostic testing may be performed on peripheral blood.
Hodgkin lymphoma	Slight male predominance in ages 5–14	15–19 years	Laboratory tests (ESR, CBC, renal and liver function, albumin, LDH, alkaline phosphatase), radiographic imaging (chest X-ray, CT, MRI, PET), bone marrow biopsy, selective biopsy of other sites. Nonspecific inflammatory markers (CRP, copper, ferritin) are not diagnostic, but helpful in monitoring.
Non-Hodgkin lymphoma	Boys > girls	Varies based on subtype	Laboratory tests (CBC, electrolytes, LDH), radiographic imaging (CT, PET), biopsy.

CBC = complete blood count; CRP = C-reactive protein; CT = computed tomography; ESR = erythrocyte sedimentation rate; LDH = lactate dehydrogenase; MRI = magnetic resonance imaging; PET = positron emission tomography.

Table 2. Manifestations of Leukemic Organ System Involvement[21-23]

Organ System	Clinical Manifestations
Central nervous system	Increased intracranial pressure (vomiting, headache, papilledema, lethargy), seizures, central nerve palsies, nuchal rigidity
Gastrointestinal system	Hepatosplenomegaly secondary to infiltration, hepatic dysfunction
Genitourinary tract	Testicular enlargement (unilateral or bilateral), enlarged kidneys secondary to infiltration of leukemia, priapism (rare)
Integumentary	Infiltration of leukemic cells causing a purplish discoloration called leukemia cutis, petechiae, and bruising
Musculoskeletal system	Bone pain, joint pain, limp (secondary to leukemic infiltration, expansion of the marrow by leukemic cells, or bone infarction), osteopenia, fracture, vertebral compression
Ocular	Retinal hemorrhages, ocular motor palsies, changes in visual acuity (secondary to leukemic infiltration of the optic nerve), photophobia, pain, blurring, decreased vision

More than one-third of children with leukemia present with fatigue, fever, bruising, bleeding, pallor, lymphadenopathy, hepatomegaly, splenomegaly, infections, rash, limb pain, limp, rib pain, or back pain.[21, 22] Additional symptoms may present based on the organ systems with leukemic infiltration (see Table 2[21-23]).

ALL accounts for about 70% of the new cases of childhood leukemia in the United States each year.[2] According to data from the NCI's Surveillance, Epidemiology, and End Results (SEER) program, the five-year relative survival rate for ALL among people diagnosed under age 45 (all races, both sexes) is 80.3%.[8]

Diagnosis. To diagnose a child with ALL, bone marrow aspirates and biopsies are often obtained from the child's posterior superior iliac crest; many children receive an anesthetic or sedative prior to this brief procedure. If bone marrow morphology reveals more than 25% blasts, ALL is diagnosed.[9] (To learn the definitions of common cancer-related terminology, see *Terms Commonly Used in Hematologic Childhood Cancers*.)

Treatment. When children are diagnosed with ALL, several variables are used to determine their disease-associated risks and the appropriate treatment regimen. The following features, which are associated with higher-risk disease, require more intensive combination chemotherapy regimens[10, 24]:

- age under one year or over 10 years
- involvement of sanctuary sites (extramedullary sites that are difficult to penetrate with chemotherapy), such as the central nervous system (CNS) or testes
- initial white blood cell count greater than 50,000/microliter
- failure to achieve remission or the presence of minimal residual disease after induction therapy

Other treatment considerations for children with ALL include genetic, cytogenetic, and biological features, such as having high hyperdiploidy (more than 50 chromosomes), which is associated with an excellent prognosis; hypodiploidy (fewer than 44 chromosomes), which is associated with a poorer prognosis; and various chromosomal rearrangements, some of which may be used to determine optimal therapy.[10, 24]

Induction therapy. The first four to six weeks of pediatric ALL therapy is known as the induction phase. Treatment is intensive: several different chemotherapeutic agents are administered, with the goal of achieving complete remission. Although subsequent therapy is required to prevent relapse, the time required to eliminate the majority of leukemic cells is the most reliable prognostic factor in ALL.[10, 24] Nursing care during this period focuses on reducing risk of infection and other complications.

Consolidation therapy, which follows induction, seeks to eliminate residual disease that remains after complete remission. The consolidation phase lasts between six and nine months, depending on the patient's level of risk and the protocol used. To minimize the risk of drug resistance, drugs that were used during induction are generally avoided during this phase of treatment.[24]

Maintenance therapy follows the consolidation phase. Its duration is about 24 months for girls and as many as 36 months for boys, who may receive a longer course in some protocols because of the risk of testicular relapse.[24]

Children with CNS involvement require treatment by intrathecal chemotherapy, systemic administration of chemotherapeutic agents that cross the blood–brain barrier, or CNS radiation, though the latter is avoided if possible because of the detrimental effects on growth and neurocognition, as well as the risk of secondary CNS tumors.[10, 24]

CNS prophylaxis. Even children with ALL who have no clinical signs of CNS disease require intrathecal prophylaxis chemotherapy to prevent progression to CNS involvement.[24]

HSCT. In patients with very advanced or high-risk ALL, allogeneic HSCT may be considered.[10,24]

AML affects far fewer children than ALL. It represents about 18% of new childhood leukemia diagnoses in the United States each year.[2] According to data from the NCI's SEER program, the five-year relative survival rate for AML among people diagnosed under age 45 (all races, both sexes) is 59.5%.[8]

Diagnosis. A child is often diagnosed with AML after the disruption in hematopoiesis results in neutropenia, anemia, and thrombocytopenia. Presentation, however, may vary considerably among children with AML. For example, those with a variation called acute promyelocytic leukemia commonly present with severe bleeding and are at risk for fatal hemorrhage.[11] Bone marrow aspirates are obtained for morphology, although if the child's condition contraindicates bone marrow aspiration, initial diagnostic testing may be performed on peripheral blood.[11] Between 20% and 25% of children with AML develop extramedullary leukemia, in which chloromas (tumor nodules consisting of leukemic cells) form outside the bone marrow, infiltrating the skin, CNS, gingiva, liver, spleen, or testes. Extramedullary leukemia may be the first sign of disease in infants with monocytic AML.[21] Additional manifestations, especially related to a high white blood cell count, may cause oncologic emergencies.

Treatment. As with ALL, treatment of AML is based on risk stratification, in which variables such as chromosomal rearrangements are relevant, and begins with induction chemotherapy.[11,25] As many as 70% of children undergoing AML therapy develop bacterial infections and about 20% develop invasive fungal infections. As prophylaxis against such infections, antimicrobial and antifungal medications are often administered.[11,25,26] Standard precautions to reduce the risk of hospital-acquired infections should be observed. These are generally driven by institutional protocols and may include the following[27]:

- ensuring that all children treated for cancer receive annual influenza immunizations in accordance with CDC guidelines
- rigorously enforcing hand disinfection and safe food handling practices
- encouraging diligent oral hygiene
- controlling the number of visitors and limiting visitation to healthy contacts
- using laminar air flow with high-efficiency particulate air (HEPA) filtration

Consolidation and maintenance therapy. Following induction chemotherapy, practices vary as to whether and to what degree patients will receive cycles of consolidation and maintenance chemotherapy. In some cases, this is based on the "quality of remission" following induction.[26]

HSCT. Both autologous and allogeneic HSCT have been investigated as postremission consolidation therapy in childhood AML, but autologous HSCT has shown no benefit. Given the toxicity associated with its requisite conditioning regimen, autologous HSCT is used only rarely in childhood AML—for example, in cases of relapsed acute promyelocytic leukemia with no detectable minimal residual disease.[11] Allogeneic HSCT, on the other hand, may be included in the treatment regimen of certain patients, depending on their risk stratification and the presence of disease following induction therapy.[11,25] In high-risk patients, allogeneic HSCT may give patients a new hematopoietic and immunologic system that can help them maintain remission, a phenomenon called the "graft-versus-leukemia effect."[11,28] Its use in first remission, however, is controversial.[11,25,26]

Terms Commonly Used in Hematologic Childhood Cancers

Blasts: immature cells

Cytogenetics: the study of heredity and variation that employs both cytology and genetics

Febrile neutropenia: a fever in the setting of neutropenia (an absolute neutrophil count ≤ 500 cells/mm^3, or a count < 1,000 cells/mm^3 that is expected to drop to < 500 cells/mm^3) that is determined by a single oral temperature measurement at or above 38.3°C (100.94°F) OR a temperature at or above 38°C (100.4°F) sustained for at least one hour.

Histopathology: microscopic examination of tissue by a pathologist to aid in diagnosis

Immunophenotype: the immunochemical and immunohistologic characteristics of a cell or group of cells

Intrathecal chemotherapy: chemotherapy injected into the spinal column during a lumbar puncture to treat or prevent disease in the central nervous system

Minimal residual disease: the presence of leukemic cells detected by flow cytometry or polymerase chain reaction laboratory evaluations in the absence of any clinical or microscopic evidence of disease

Molecular genetics: the study of the structure and function of genes at a molecular level

Morphology: microscopic examination of blood cells to evaluate their characteristics and determine the presence of disease

Remission: no clinical evidence of the disease

As with ALL, CNS treatment is given—usually by intrathecal chemotherapy or systemic administration of chemotherapeutic agents that cross the blood–brain barrier—to all children with AML, including those with no detectable CNS involvement, to whom it is given as prophylaxis.[11, 25, 29]

CHILDHOOD LYMPHOMA

Lymphoma is the second most common hematologic childhood cancer diagnosed in U.S. children and adolescents under age 20.[2] Lymphoma occurs when lymphoid cells become malignant. Although lymphoma usually involves the lymph nodes, it may also originate in primary lymphoid tissues, such as the bone marrow and thymus, or in secondary lymphoid tissues, such as the spleen, mucosa-associated lymphoid tissue, or nonlymphoid organs.[18] Pediatric lymphoma is categorized as either Hodgkin lymphoma or non-Hodgkin lymphoma (NHL), with various subtypes in each category.[30]

Hodgkin lymphoma is the most common malignancy in American adolescents, accounting for approximately 3% of cancer diagnoses in children up to age 14 and approximately 13% of cancer diagnoses in adolescents between the ages of 15 and 19.[2] November 2017 SEER data show the five-year relative survival rate for Hodgkin lymphoma among people diagnosed under age 45 (all races, both sexes) to be 93.9%.[8]

Diagnosis. Children and adolescents with Hodgkin lymphoma may present with lymphadenopathy, often located in the supraclavicular or cervical area.[18] Additional symptoms depend on disease location. For example, tumors in the chest may cause dyspnea, cough, hypoxia, and pleural or pericardial effusion, while tumors in the abdomen may cause hepatomegaly or splenomegaly.[18] Certain symptoms, termed "B symptoms," are consistent with systemic disease and poorer prognosis, whether seen independently or in combination. These include the following[18]:

- fever of at least 38°C (100.4°F) for three consecutive days
- unexplained weight loss of at least 10% body weight over a six-month period
- drenching night sweats

Patients with suspected Hodgkin lymphoma undergo physical examination, radiographic imaging, and biopsy of specific sites, as well as laboratory testing for nonspecific inflammatory markers that are not diagnostic but are helpful in evaluating and monitoring the patient.[12]

The Ann Arbor Staging Classification for Hodgkin Lymphoma is used to classify the clinical stages of Hodgkin lymphoma.[31] This system describes the location of involved lymph nodes, the number of sites involved, whether they are on one or both sides of the diaphragm, and the presence of any organ involvement, with stage IV describing the most diffuse disease or involvement of the liver, bone marrow, lungs, or CNS.[31] Within each stage, the designation of category A or B indicates the absence or presence of B symptoms, respectively; designation of category E signifies a single extranodal site and S signifies splenic involvement.[31]

Treatment. Several treatment regimens exist for Hodgkin lymphoma, each containing multiple agents with varying toxicity profiles so that doses can be optimized. Protocols are determined based on the disease presentation and response to initial therapy. Although radiation has been shown to be very effective against the disease, it is avoided when possible owing to the associated risks of cardiovascular disease, thyroid dysfunction, and secondary cancers.[32] Relapsed Hodgkin lymphoma has been reported to occur in approximately 16% of patients[33] and requires additional therapy, possibly including monoclonal antibodies[34, 35] and autologous HSCT.[18, 36]

Nursing care of the child with cancer often requires the administration of blood products, including platelets and packed red blood cells, to overcome treatment-related myelosuppression.

NHL is responsible for nearly 7% of childhood cancers in U.S. patients under age 20.[2] NHL includes several types of lymphoma, which vary in incidence and frequency based on the patient's age at diagnosis.[13] The most common types of NHL in children and adolescents are Burkitt lymphoma, diffuse large B-cell lymphoma, lymphoblastic lymphoma, and anaplastic large-cell lymphoma.[30]

Diagnosis. As with Hodgkin lymphoma, painless lymphadenopathy may be a common presenting symptom of NHL, but the spread of disease is more rapid.[12] Additional symptoms are based on the location of the tumor and may include abdominal manifestations such as bowel obstruction, respiratory distress secondary to a mediastinal mass, or neurologic dysfunction secondary to CNS involvement.[12] As in Hodgkin lymphoma, patients with suspected NHL undergo physical

examination, radiographic imaging, biopsy of specific sites, and laboratory testing, including for lactate dehydrogenase, complete blood count, and electrolyte levels. In the absence of a clinical history and examination findings suggestive of malignancy, patients do not require immediate biopsy but are reexamined within a month. Surgical diagnostic biopsy is generally performed only on children with chronic or generalized lymphadenopathy or symptoms of new-onset systemic disease.[18]

Classification of NHL in children follows the St. Jude staging system, which classifies disease from I (less severe disease) to IV (more severe or extensive disease). According to these criteria, bone marrow or CNS involvement are classified as stage IV and primary intrathoracic tumors or extensive abdominal disease are classified as stage III.[12] Recently, there has been interest in revising the criteria to include other important characteristics of the disease, such as cytogenic, molecular, and immunophenotypic features, which may affect prognosis.[37]

Treatment. Systemic, multiagent chemotherapy regimens are the mainstay of treatment in NHL.[13] Monoclonal antibodies have been successful in treating adults with NHL and are increasingly used in pediatric regimens as well. Regimens are selected based on the type of NHL and histologic findings.[13] Surgery is not a treatment of choice in lymphoma. Use of radiation therapy is limited to cases of CNS involvement in older children and such oncologic emergencies as airway compromise, spinal cord compression, or superior vena cava syndrome secondary to compression caused by the tumor.[18] Long-term survival rates vary significantly based on the subtype and stage of NHL, but range from 80% to 90% for most subtypes. For advanced anaplastic large cell lymphoma, however, long-term survival rates range from 60% to 75%.[38] As with Hodgkin lymphoma, advanced, refractory, or relapsed NHL may be treated with autologous HSCT. Patients with certain NHL histologic subtypes (those for which the presumed "graft-versus-lymphoma" effect is expected to be beneficial) may receive allogeneic HSCT,[39] although this is usually considered only after an autologous transplant.

THE FAMILY'S NEEDS

A diagnosis of childhood cancer is a stressful event. It takes time for families to emotionally process the diagnosis before they can learn how to care for their child as therapy is initiated and a discharge plan is developed. An interprofessional expert panel convened by the Children's Oncology Group recommends that family and patient teaching be provided across the continuum of care at a pace that addresses the most immediate needs first and does not overwhelm the family.[40-42] Nurses, who find themselves at the bedside during the family's most vulnerable and difficult times, play an instrumental role in providing the education and anticipatory guidance required to help families successfully transition back to their homes. Depending on the diagnosis and response to therapy, a child may continue receiving cancer treatment for months to years; and even when treatment is completed, the supportive care nurses provide regarding ongoing monitoring and follow-up is invaluable to families navigating their "new normal."

One challenge of understanding pain in children is that parents or caregivers often rate children's pain at a lower level than the children do.

SUPPORTIVE CARE

Treating pain. Pain is one of the most frequently reported symptoms in children with cancer. Pain may be related to the cancer itself, or associated with therapy or procedures related to cancer treatment.[43,44] One challenge of understanding pain in children is that parents or caregivers often rate children's pain at a lower level than the children do.[44] Because of the impact of pain on quality of life in children and young adults, it is critical that nurses assess pain regularly, using self-report when possible, so they can advocate for appropriate interventions. Effective pain management may be achieved through both nonpharmacological therapies, such as distraction, relaxation, massage, or yoga, and pharmacologic intervention.[43]

Meticulous mouth care. Nurses need to pay specific attention to the patient's oral care during treatment, documenting the occurrence of mouth sores and mucosal breakdown (mucositis). Saline rinses, regular brushing with a soft toothbrush, and flossing when it will not cause injury or bleeding should be reinforced during hospitalization to promote continuation of these routines after discharge to home. When severe mucositis is evident, foam or very soft

brushes may be substituted for toothbrushes[27] and analgesics may be administered as needed.[45]

Prevention and treatment of nausea and vomiting. Scheduled or as-needed medications may be administered for chemotherapy-induced nausea and vomiting. Nurses play an important role in advocating for antiemetic medication orders and can let providers know the efficacy of various agents and regimens in individual patients. Nurses can also support families in using such nonpharmacological interventions as relaxation training, aromatherapy, and distraction,[46] collaborating with pharmacy colleagues to ensure that herbs or other remedies requested by families will not harm the patient or interact with the therapy regimen.

Transfusion support. Nursing care of the child with cancer often requires the administration of blood products, including platelets and packed red blood cells, to overcome treatment-related myelosuppression. It's important to monitor patients closely throughout the transfusion and after its completion for signs or symptoms of adverse reactions.

Care of central venous catheters. Many pediatric institutions participate in quality improvement initiatives to reduce the incidence of bloodstream infections through bundled care approaches. Nurses should be aware of their hospital protocol and strive to implement evidence-based practices related to catheter care. Research supports the use of chlorhexidine gluconate baths to reduce the risk of central line–associated bloodstream infections and hospital-acquired infections as a part of catheter maintenance care.[47-49] This practice has been integrated into many central line care bundles.

Advocacy for supportive therapies. These may include therapies provided by certified child life specialists, who help children and their family members adjust to the disease and its treatments through developmentally appropriate patient teaching, treatment preparation and rehearsal, medical play sessions, and procedural support such as individualized music playlists or audiobooks. There is evidence that integration of the supportive therapies child life specialists introduce can reduce the frequency of daily anesthesia children with cancer require when treated with radiation therapy.[50] Children with cancer may also benefit from working with registered music therapists, as studies have shown that children provided with music therapy before, during, and after a painful procedure report significantly lower pain scores and anxiety levels than those given usual care.[51] Nurses are uniquely positioned to recognize distress in children and families and to advocate for such therapeutic interventions. Social workers and case managers or care coordinators can assist families with insurance and other practical issues related to the care of their child.

Chemotherapeutic agent–specific care. Depending on a child's treatment protocol, nurses may be required to provide additional care and monitoring, such as

- hydration support with monitoring by urine specific gravity.
- administration of analgesics and other supportive therapies.
- performance of special skin care in accordance with institutional protocol.

PATIENT AND FAMILY EDUCATION

Information about diagnosis and treatment. It's important to teach families about the body systems affected by the cancer and to explain the potential adverse effects of treatment, particularly those related to the digestive and hematopoietic (bone marrow) systems, which are the systems most frequently involved. Families should feel prepared to manage such adverse effects as nausea and must know the necessary precautions to take when the child is neutropenic or thrombocytopenic.[41]

Nurses need to pay specific attention to the patient's oral care during treatment, documenting the occurrence of mouth sores and mucosal breakdown.

Care of an external central venous catheter. Children discharged to home with external central venous catheters require routine catheter care, which is generally administered by their family. Care includes flushes to maintain patency of the line and regular dressing changes. During the child's hospital stay, nurses should provide family members with essential care instruction and supervised practice sessions. Patient and family teaching should include information on

- emergency treatment if the catheter were to break or become dislodged.

- bathing precautions: the catheter must be completely covered and kept dry during bathing, and swimming is prohibited owing to risk of infection.
- chlorhexidine use: to reduce risk of infection, chlorhexidine should be used for skin antisepsis during dressing changes and for baths or showers in accordance with institutional policy.[47-49]

tional protocols; neutropenic fever is an oncologic emergency warranting immediate intervention.[40,41] Generally, temperature is measured once or twice daily and if the child feels ill or is warm to the touch. Family members should be advised that fever, which has been variously defined as a single oral temperature measurement at or above 38.3°C (100.94°F),[53] a temperature at or above 38°C (100.4°F) for at least one hour,[53] and a single temperature above 38.5°C (101.3°F),[54] may warrant an immediate trip to the ED and notification of the oncology team. Nurses should ensure that family members and caregivers know the institutional standards of their child's treatment center. Febrile patients require a workup for infection, including blood cultures, and intervention with empiric antibiotic therapy. Additional therapy may be warranted based on the patient's clinical condition.

Infection is one of the most common complications of childhood cancer therapy. Neutropenic fever is an oncologic emergency warranting immediate intervention.

Myelosuppression puts children at risk for bleeding and bruising. During myelosuppressive periods, parents need to make sure that their children avoid contact play, instead engaging in quiet, low-impact activities. Parents should be advised that shortness of breath, dizziness, or pale color (especially of the mucous membranes) should prompt a call to the child's oncologist, as these signs may indicate anemia and the need for transfusion.

Handwashing is the most important intervention to prevent infection in families who are caring for a child with cancer. While alcohol-based hand rubs are an acceptable substitute for handwashing in most cases, soap and water is indicated in the setting of infectious diarrhea or when hands are visibly soiled.[27,41]

Safe food handling. Although special diets designed to reduce bacterial contamination are not supported by evidence,[27] it is important to teach families about safe food handling, the importance of refraining from eating raw or uncooked food, and precautions to take when eating out. For example, families should[52]

- avoid eating from buffets and salad bars, where foods are likely to sit out for long periods and be exposed to many people.
- ask the server to let them pack their own leftovers and refrigerate them as soon as possible.
- choose restaurants that are clean, and request that they prepare food as ordered and that the food is fresh at the time it is ordered.

The U.S. Department of Agriculture (USDA) and Food and Drug Administration (FDA) have published the booklet *Food Safety for People with Cancer*, which is available for download at www.fda.gov/media/83710/download.

Responding to fever. Infection is one of the most common complications of childhood cancer therapy. When a child is neutropenic, it is important to monitor the child's temperature in accordance with institu-

Medication administration. Hospitalization provides an important opportunity to demonstrate medication administration techniques and reinforce the indications for each medication administered. Nurses need to explain storage recommendations and encourage strict adherence to medication regimens to promote optimal outcomes.

Integration of palliative care services upon diagnosis is consistent with the Institute of Medicine's recommendation that "palliative care can begin early in the course of treatment for any serious illness."[55] Nurses can introduce and advocate for palliative care services and inform families of the goal of preserving and improving quality of life while managing symptoms.

COMPLETING TREATMENT

Despite the success and relief associated with completing treatment, families have reported challenges associated with this transitional time. They have described feeling happy and grateful to be completing therapy, but lost and uncertain during the transition.[56] Nurses can help ease the family's transition from the frequent visits with the health care teams required during therapy to the anticipatory supportive guidance they will receive as they move toward cancer survivorship care.

After a prolonged period of intensive care provided almost exclusively by the oncology team,

families may be reluctant to transition back to a primary care model or cancer survivorship clinic. As childhood cancer survivors complete treatment and approach adulthood, however, their needs are better met in survivorship clinics. Nurses are in a unique position to reinforce the importance of monitoring patients for symptoms related to the underlying disease or late effects of therapy for months to years following completion of treatment. Nurses are also well positioned to support the collaborative relationship between primary and specialty care upon treatment completion, while identifying resources for off-treatment patients.

Necessary follow-up. Survivors of childhood cancer require regular follow-up and monitoring for late effects of therapy. Complications of treatment may affect various organ systems, producing pulmonary, gastrointestinal, cardiovascular, musculoskeletal, reproductive, endocrine, auditory, ocular, oral, dermatologic, urinary, neurologic, immunologic, and psychological symptoms.[57] When evaluating patients for late effects of treatment, it's necessary to consider the specific therapies that each patient received. A publication of the Children's Oncology Group, *Long-Term Follow-Up Guidelines for Survivors of Childhood, Adolescent, and Young Adult Cancers*, includes potential late effects associated with each treatment as well as recommended evaluations and counseling for the patient and family.[58] ▼

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Jessica Lynne Spruit is an assistant clinical professor and specialty coordinator of the Pediatric Acute Care Nurse Practitioner program at Wayne State University, and a pediatric NP in the Division of Hematology/Oncology at Children's Hospital of Michigan, both in Detroit. Contact author: jessica.spruit@wayne.edu. *The author and planners have disclosed no potential conflicts of interest, financial or otherwise. A podcast with the author is available at www.ajnonline.com.*

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