PART I
FOUNDATIONS
INTRODUCTION

“...I believe there is merit in looking at the past in order to have a perspective of accomplishments, challenges, trends and future needs. Moreover, I think it can give us courage and determination to pursue our ideas and ideals.”

The history of pediatric hematology and oncology nursing as distinct subspecialties began in the 1940s. The complex factors prompting the emergence of these specialties are rooted in the histories of hematology, oncology, pediatrics, and nursing. This chapter describes the mosaic of traditions, beliefs, values, and practices that influenced the past, shape the present, and suggest the future of pediatric hematology/oncology nursing. This chapter also describes ways in which historical forces continue to shape attitudes and behaviors regarding these specialties.

WERE BLOOD DISORDERS IN CHILDREN RECOGNIZED THROUGHOUT HISTORY?

Little was known about disorders of the blood in adults or children until after the Renaissance. Information primarily concerned the assumptions, meanings, and attitudes about blood—a substance at once common and easily visible yet a mystery.

Blood was recognized as an essential, vital fluid that was needed to sustain life centuries before its circulation, roles, and functions were established. In the Iliad, Homer related that Asclepius, the god of medicine, possessed the ability to stop bleeding. Hippocrates identified blood as one of the four humors that governed health and illness. There are more than 500 references to blood in the Bible, including Leviticus 17:11, “The life of the flesh is in the blood.”

With rare exceptions, validation of the presence of diseases of the blood has come from paleo-pathology investigations of ancient skeletal remains and mummies. These studies indicate that hematologic disorders have existed for centuries. Exactly what those conditions were and what caused them was not always clear. For example, skeletal changes in the skull consistent with prolonged bone marrow hyperplasia indicate long-duration severe anemia. Such findings, noted in adult and child skeletons from several countries, could be the result of nutritional disorders, malaria, or chronic blood loss, as well as distinct hematologic disorders such as thalassemia and congenital hemolytic anemia.

Some writings indicated awareness of blood-related conditions. The Babylonian Talmud, for example, lifted the obligation of infant circumcision if two successive sons of one mother or sons of two sisters within the same family did not survive the ritual. This suggestion of the possibility of a familial bleeding disorder is considered the earliest reference to hemophilia.

For centuries, hematology problems in children received little attention because these disorders appeared to be uncommon, few diagnostic tools were available to identify abnormalities, and no adequate treatments existed. The development of the microscope in the 17th century permitted visualization of blood cells, but its impact was minimal for almost a century. Even as the microscope improved in magnification, many clinicians of the time viewed it as an apparatus that took away from—rather than enhanced—clinical observation and decision making.

The first scientific papers devoted to pediatric hematology concentrated on neonates—an age group of focus for decades. Georges Hayem’s Du Sang et de ses Altérations Anatomiques published in Paris in 1889 described the number, type, and structure of neonatal blood cells he had identified. It took decades to quantify infant blood volume, cellular composition, and possible relationships to neonatal jaundice. Neither of the first two pediatric hematology texts, published in Europe in 1925 and 1928, defined red cell survival times and blood volume measurements. In the United States, Luther Emmett Holt’s seminal text, The Diseases of Infancy and Childhood (1897), briefly discussed hemorrhagic disease of
newborns and selected other disorders. William Palmer Lucas and E. C. Fleischner contributed chapters on blood to the first edition of *Abt’s Pediatrics* (1924). These chapters are considered the first American comprehensive publication on pediatric hematology. Kenneth Blackfan and Louis Diamond’s *Atlas of the Blood in Children* (1944) featured the then-known blood values of children and color depictions of blood cells. Carl Smith’s *Blood Diseases of Infancy and Childhood* (1960), the first American pediatric hematology text, and Nathan and Oski’s *Hematology of Infancy and Childhood* (1974) made significant contributions to advancing the field.10,11 Additional selected hematology milestones are noted in Table 1-1.5,8-16

Remarkable progress has been made since the description of the first pediatric hematology condition by Rudolf von Jaksch in 1889,9 but the pace of advancement has been variable. It took until the late 1940s to develop the techniques of bone marrow aspiration9,17 and to establish marrow characteristics in a variety of diseases. Understanding normal blood values among different age groups (a prerequisite for being able to define abnormalities) was not achieved fully until the 1950s.10 Refinements in diagnosis required advances in immunology and genetics.

**HAS PEDIATRIC CANCER ALWAYS EXISTED?**

Hippocrates (460-370 BCE) was the first to use the term *karkinos*, or crab, to describe a cancer. It was an imprecise term describing a variety of conditions, some of which correspond to the present definition of malignant neoplasm.18 Hippocrates mentioned neck growths in children, but it is not known whether these growths were cancers.19 Swellings often were considered a cancer if the patient died.20 Although true malignancies did exist during this time, paleo-pathology studies limited by diagnostic technology and sample size suggest that cancer was an uncommon occurrence.20,23

For centuries, cancer in children was a rare, poorly understood health problem that often was misdiagnosed. In 1809 James Wardrop described 24 cases of malignant eye tumors, 20 of which occurred in children younger than 12 years of age. This report represented the first collection of childhood cancer cases.24 In 1876 C. J. Duzan published a tabulation of 182 pediatric malignancies reported in the literature between 1832 and 1875. Duzan’s work represented the first publication devoted exclusively to cancer in children.19

Determining the incidence and prevalence of childhood malignancies has been a daunting task. Procedures for collecting and reporting cancer statistics were not clarified and organized until the 20th century. In the United States, for example, cancer mortality for all people younger than 30 years of age was presented as a single-age category. James Ewing, the pathologist for whom Ewing sarcoma is named, objected strongly to that reporting system. He believed that juvenile cancers “...are so peculiar that properly they may not be compared with any adult tumors, and that this entire subject deserves to be treated as a special department in the descriptive history of neoplastic disease.”25(p14) It is now understood that cancers of adolescents and young adults also require particular focus.

In the United States various government agencies have collected census data and other vital statistics since the late 1800s. Much of this historical information is available through the Annual Reports of U.S. Vital Statistics in print or online. Definitions of vital statistics were clarified over the years. Because pediatric cancers were not reportable diseases and state tumor registries were few in number, basic reporting data were unavailable or incomplete. This problem was exacerbated because leukemia and Hodgkin lymphoma were classified as separate from pediatric cancers.24

It took until the end of World War II for the problem of pediatric cancer to be in the words of Farber, “unmasked”.26 Pediatric cancers became the leading cause of death from disease among children as advances in general pediatrics, particularly the control of infectious diseases, changed the landscape of pediatric mortality. Malignancies continue to be the leading cause of death from disease in the 1–19-year-old age groups27 (Table 1-227).

**WHAT CAUSES CANCER?**

Throughout the centuries and across cultures, mankind has struggled to understand the complex entity known as cancer.28 Theories suggested in each era reflected the way in which diseases were understood at that time. Although some theories seem outlandish, ill-conceived, or even
PART II
TREATMENT
INTRODUCTION

Most families who have children with cancer agree that the period of diagnosis is one of the most difficult times of their cancer journey. The nurse caring for the child suspected of having a malignancy is in a unique position to offer support and guidance at this stressful time. To do this, however, the nurse needs a thorough understanding of the various measures used to diagnose pediatric malignancies and their usual sequence. Preparing the child and family for tests and procedures and facilitating patient safety are key nursing responsibilities.

ESTABLISHING THE DIAGNOSIS

Although cancer is the leading cause of death from disease in children 1 to 19 years of age in the United States, it is nevertheless uncommon. The diagnosis of childhood cancer is often delayed, despite a family’s persistence in returning repeatedly with their child to see the family doctor or pediatrician. The initial symptoms of childhood cancer can mimic benign conditions such as infection or injury. Many children are treated with several courses of antibiotics for persistent infectious symptoms or visit subspecialists for assessment of a specific pain or organ dysfunction.

The signs and symptoms of cancer are related to the age of the patient, the type of tumor, and the extent of the disease. The greatest aid in diagnosing malignant disease in children is a high index of suspicion, because there is no classic, universal symptom or symptom complex for cancer in children. A recent review of 25 studies investigating delays in diagnosis of childhood cancer showed that the mean “delay time” varied by cancer type from a low of 2.5 weeks for children with Wilms tumor to a high of 29.3 weeks for children with a brain tumor. An Israeli study showed an average of 15.8 weeks’ delay with a range of 0 to 208 weeks for all childhood cancers. In the case of brain tumors, a Swedish study found a median delay of 9 weeks with a range of 1 to 19 weeks.

The goals of the diagnostic and staging phases of treatment are to determine the presence of a cancer, to identify its type and stage, and to identify its location(s) throughout the body. A thorough evaluation of baseline organ function is undertaken for the child who will undergo cancer therapy. These procedures are carried out expeditiously to allow appropriate therapy to begin as quickly as possible.

New and sophisticated tests that provide detailed information and prognostic data are available only at established pediatric cancer centers. As the science of pediatric oncology advances, classification of tumors and leukemia has become more precise and complex. Treatment is often stratified by these classifications, resulting in therapy that is tailored to the individual. Treatment may be aggressive for some but less intensive for others. Every child deserves state-of-the-art therapy and the psychosocial support that a team of professionals experienced in the care of children with cancer can provide. Childhood cancer survival rates are positively influenced by the place of treatment and the use of cancer protocols. Further studies have demonstrated the importance and significant survival advantage to children and adolescents when their treatment is based on well-defined protocols in specialized children’s cancer centers.

Basic, noninvasive imaging procedures that confirm the presence of a mass and/or basic laboratory work suggesting organ dysfunction are often performed by the primary healthcare provider. Based on the results, a prompt referral is made to a pediatric oncologist. Extensive community-based diagnostic testing should not be pursued, as many of the diagnostic tools required are unavailable in the community. The lack of a detailed and comprehensive diagnostic evaluation subjects the child and family to unnecessary delay, expense, and trauma.

Patient History

Diagnosis begins by obtaining a detailed medical history. Special attention should be given to
factors that suggest the possibility of malignancy. The interview is best conducted privately, without interruptions, and with consideration for the comfort of both the parents and the child. The parents will be more relaxed during the history-taking process if adequate provision has been made for the child’s needs. An ill child should be assigned to the care of an extended family member or a staff member. A more active child may be provided with toys or other activities. The parent may prefer to hold an infant or an older child, or the child may be placed on the examination table close to the parent.

Begin the interview process by greeting both the parent and the child. Including a friendly comment to young children about their appearance, clothing, or toys may help engage them. During the interview, make frequent eye contact with the child. Every attempt should be made to engage the child verbally during anxiety-provoking moments.

The older child and adolescent should actively participate in the initial history taking, as this sets the expectation of their involvement throughout their course of their care. Give the adolescent the opportunity to provide information without his or her parents in the room, usually after the initial interview with the parents and before the physical examination. The parents can be reassured that the pertinent findings of the physical examination will be shared with them. Respecting the teen’s independence and establishing a sense of confidentiality are best achieved if begun at the onset of treatment.9,10 Of course, if the adolescent is frightened and prefers to have the parents present, that wish should be respected.

To collect relevant information, the examiner should phrase questions in a way that the parent and adolescent can easily understand, listen carefully to the responses, and encourage the family to express ideas and concerns freely. If the parents are not fluent in English, an interpreter with some degree of medical knowledge should be obtained. The availability of trained translators is required by the Joint Commission.11 Relying on untrained translators, such as bilingual children or other family members who are trying to absorb information and transmit it while emotionally upset, is inappropriate.12

The examiner elicits the information by following a specific pattern. First, determine the chief complaint. Many health professionals ask the parents, “Why did you come to see us today?” The examiner then develops the sequence of the present illness by inquiring about the date at which symptoms first appeared, the order of their occurrence, the diagnoses made by the referring healthcare provider who examined the patient, and the response to any prescribed treatment. Listen intently to the parent’s description of the child’s illness or complaint. Parents describe their child’s problem as they perceive it, and their story frequently includes a theory regarding the cause of the problem. Understanding such theories may be helpful later when counseling parents.13

The examiner next reviews the child’s history. The child’s prenatal, neonatal, and subsequent growth and development are essential information. Children with constitutional chromosomal abnormalities have an increased risk of malignancy. Any history of immunodeficient or metabolic diseases or genetic disorders such as autoimmune diseases, neurofibromatosis, and Down syndrome is documented.14 Record all immunizations and past illnesses and obtain a thorough social history. A routine pediatric social history includes age, marital status, and occupation of the parents—including stepparents or those with whom the child lives—and the ages of siblings. The parent or the older child is asked to discuss school performance and adjustment. Information about the family’s financial status, including insurance coverage, is obtained. Inquire about the family’s housing situation and available social and community resources.

Pertinent family medical history is also documented. Any history of cancer in family members, including the health status of grandparents, parents, and siblings, is noted, with special attention given to any pediatric cancers or indication of familial cancer syndromes (as discussed in Chapter 20). A family pedigree is constructed and documented on the medical record if there are several family members with cancer histories.

Finally, the patient history concludes with a review of body systems. The examiner attempts to elicit any symptoms that the parents may not have recognized or considered relevant.
Questions are asked about each body system. Before concluding the interview, the examiner asks the parents if all their questions and concerns have been discussed. No concern should be minimized or automatically dismissed.

**Physical Examination**

The physical examination (Figure 7-1) begins when the examiner observes the child while obtaining the medical history. Initial impressions about the child-parent interaction, the child’s general appearance, and whether the child behaves in an age-appropriate manner are formed. A developmental assessment is an integral part of a pediatric physical examination. During the physical examination there must be regard for the child’s privacy and comfort. The child should be treated with respect and consideration. An infant or toddler can be examined almost completely with the child on the parent’s lap.

During the physical examination, adolescents may raise concerns about their health or their bodies that were not mentioned in the initial interview. Many teens have misguided ideas about the cause of their illness, the extent of their symptoms, and the prognosis of childhood cancer. The skilled practitioner should convey to the adolescent that he or she is the most important concern of the medical team. The teen needs to be the primary source of information and is encouraged to be active and involved throughout treatment and decision making.

Vital signs are obtained and recorded during the initial examination. Height and weight without shoes are measured and plotted on the appropriate growth chart. Metric measurements are taken and used to calculate the patient’s body surface area (BSA). The BSA is used to calculate future chemotherapy doses, except in the case of infants. Infant doses are typically calculated based on weight in kilograms, because infants have a proportionally greater BSA than older children who are larger than 10 kilograms. Care must be taken to ensure the accuracy of the height and weight measurements. Because metric units are not the standard in the U.S. and are therefore unfamiliar to many, incorrect values may be easily overlooked. Converting the metric height and weight into inches and pounds can provide a valuable double-check mechanism. The BSA is measured in square meters (ie, m²) and is often automatically computed by hospital electronic order systems. It can be computed manually by using the following formula:

\[
\text{BSA} = \frac{\text{Height (cm)} \times \text{weight (kg)}}{3600}
\]

BSA should not be confused with body mass index (BMI). The BMI is a reliable indicator of body fat in children and teens. The BMI can be calculated automatically on electronic hospital records or by using the Centers for Disease Control’s online calculator, available at http://apps.nccd.cdc.gov/dnpabmi/. After the child or teen’s BMI is obtained, the value is plotted on the BMI-for-age growth charts (for either girls or boys) to obtain a percentile ranking. The percentile indicates the relative position of the BMI among children and teens of the same sex and age. The BMI is an important calculation in pediatric oncology, as alternative formulas or adjustments of chemotherapy doses are often used for obese children.

The growth chart is an important tool for evaluating growth failure or dysfunction secondary to cancer treatment. Such dysfunction may be recognized sooner if baseline and incremental height and weight measurements are obtained and recorded regularly. The head circumference of infants and young children is also measured and recorded. The head circumference is considered the best assessment of infants’ brain growth and development. Baseline and serial
PART III
SUPPORTIVE CARE
INTRODUCTION

Pediatric oncology nurses are noted for their skilled end-of-life (EOL) care for children who are dying from disease or complications. These nurses focus on the needs of patients, families, and staff. In recent years the concept of palliative care has been introduced to improve and expand the important aspects of EOL care. Palliative care goes beyond the aspects of EOL care to provide compassionate care to children and their families at the time the child is diagnosed with a life-threatening or life-limiting illness. Palliative care has moved “upstream” from the terminal stage of illness to begin when the diagnosis of a life-threatening or life-limiting illness is made.

For pediatric hematology nurses, the palliative care role may be less clear. Many patients with hematologic disorders face many types of morbidity and early mortality. Some hematologic disorders, such as aplastic anemia, may be considered life-threatening, and palliative care principles should be incorporated into the care plan for a patient and family at the time of diagnosis. Other conditions, such as thalassemia, may be considered life-limiting, and palliative care may be involved as the disease and troublesome symptoms progress. Still others, such as sickle cell disease, may be considered chronic conditions that do not require palliative care unless or until they become life-limiting or life-threatening.

THE CONCEPT OF PALLIATIVE CARE

As defined by the National Consensus Project for Quality Palliative Care in 2009, palliative care is a dynamic process of supporting patients with debilitating and life-threatening illness and their families, regardless of the duration of the illness. Services are provided to patients of any age from the time of diagnosis until cure or death; these services include supporting families through the bereavement period. The goal is to help patients and families achieve the best possible quality of life in accordance with their values, preferences, and beliefs. Ideally, palliative care is delivered by a skilled interdisciplinary team that provides attention to pain and other distressing symptoms; emotional, spiritual, and practical support; assistance with complex medical decision-making; and coordination across the continuum of care settings.

As defined by the World Health Organization (WHO), palliative care is a broad philosophy of total, compassionate care of patients with life-threatening or life-limiting disease. The goal is achievement of the best quality of life for patients and their families by preventing and relieving suffering. Palliative care affirms life and recognizes death as a normal process. It does not hasten death, nor does it postpone it. In collaboration with families, the objectives are to prevent or relieve physical symptoms (pain as well as other distressing symptoms), maintain activity and independence for as long as comfortably possible, alleviate psychological distress (including fear, anxiety, isolation, or anger), provide for a death with as much dignity as possible, and support those who are bereaved.

Palliative care evolved from the modern hospice care movement founded by Dame Cicely Saunders in 1967. While providing a much-needed service, hospice care frequently has not met the needs of pediatric patients. First, most hospice providers focus on adult patients with life-threatening illnesses. Few, if any, staff in these hospices are prepared to deal with the medical, physiological, emotional, and developmental issues of dying children.

Second, the timing of the transition from active therapy with curative intent to hospice EOL care is more gradual in pediatric patients. Because of the higher cure rates in pediatric cancers, the objective of care, even for many high-risk patients, initially is optimistic. It may be difficult for both healthcare providers and parents to make a formal transition to non-cure-directed care. For example, parents and their child may wish to participate in a phase I study at the end
of life, but such “active” treatment patients often
do not qualify for hospice services. Families and
providers may be faced with waxing and waning
palliative care needs and require recurrent dis-
cussions over time about their changing clinical
status. These needs “do not fit neatly into the
medical, psychological, spiritual, and economic
framework established for adult EOL care.”

Too often, hospice care is not sought until the
very end of a child’s life.

Through the efforts of organizations such as
Children’s Hospice International founded by
Ann Armstrong Dailey, and advocates like Ida
Martinson, there has been increased awareness
of the needs of pediatric patients and a modest
increase in pediatric hospice services. However,
a substantial percentage of children dying of
cancer in this country still are suffering and
their symptoms are not adequately prevented
or relieved. Pediatric palliative care has been
developed to meet the needs of these patients
and their families much earlier than the EOL or
hospice stage.

PEdiatRIC PALLiative CARE

Pediatric palliative care is family-centered, ad-
dressing spiritual, social, psychological, and
physical needs. It offers children with life-
threatening illness the hope of living as actively
as possible until death, and it offers families the
support to cope during the patient’s illness and
in their bereavement.

Table 13-1 outlines some historical milestones in the development of pediatric palliative care. Some important
events in palliative and hospice care in general
have been included to illustrate the basis for
further development of pediatric care. Palliative
care specific to pediatric patients has gained mo-
momentum only during the last 2 decades with the
recognition that children and their families have
unique palliative care needs.

Several definitions of pediatric palliative care
(Table 13-2) have many similarities. The
basic principles and elements of palliative care
have been defined by Himelstein and the
National Consensus Project for Quality Palliative
Care (Table 13-3).

Although the cure rate for childhood cancer
has increased dramatically during the last 4 de-
cades, pediatric oncology nurses must approach
each child newly diagnosed with cancer with
optimistic honesty. Any diagnosis of cancer in a
child carries with it life-threatening possibilities,
especially if the expected prognosis is uncertain.
The predictability of a poor outcome in certain
childhood cancers, such as stage IV neuroblas-
toma, may be higher. No one knows at the time
diagnosis which child will live and which eventu-
ally will succumb to the illness or side effects of
therapy. Thus, pediatric oncology nurses must be
prepared to address not only the issues of survi-
vorship but also the care of terminally ill children
when death is inevitable. This includes learning
to deal with personal feelings to help children
and families traverse the terminal phase with as
much comfort, dignity, and strength as possible.
This chapter will now focus on EOL care. Keep
in mind that many of the concepts apply not only
to dying children, but also to any child with a life-
threatening illness and their families.

CHILDREN’S CONCEPTS OF DEATH

Development of Understanding

Helping children with life-threatening illness
and their families requires an understanding of
how children’s ideas and concepts about death
develop. Social, scientific, and technologic ad-
vances have produced changes that influence
a child’s experience with death. Children have
daily encounters with beginnings and endings,
separations, and losses, which all are part of the
development of the concepts related to death.

Children encounter death in a variety of ways.
In the past, when the infant mortality rate was
high and the average life expectancy was short,
a child often experienced the death of siblings
and, or parents. The modern child is less likely
to lose a parent or sibling through death. In fact,
recent generations are the first known in history
for which many middle-aged adults have not yet
experienced the death of an immediate family
member.

In addition, technologic advances and the
media have exposed children to confusing or
inaccurate concepts of death. Computers, elec-
tronic and video games, movies, and even books
designed especially for children expose children
to death. The media (television, radio, and news-
papers) presents death and the threat of death
in war or in stories of the deaths of individuals
(violent and otherwise). Even a common bed-
time prayer contains a reference to death—“If I
should die before I wake....” Rhymes and songs
may have death-related themes, fairy tales con-
<table>
<thead>
<tr>
<th>Year</th>
<th>Milestone</th>
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<tbody>
<tr>
<td>1967</td>
<td>Dame Cicely Saunders opens St. Christopher’s Hospice in London, emphasizing the multidisciplinary approach to caring for dying people with careful attention to social, spiritual, and psychological suffering among patients and families.</td>
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<tr>
<td>1974</td>
<td>Florence Wald founded the first home hospice care program in the United States; first hospices in Canada open.</td>
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<td>1976</td>
<td>Ida Martinson publishes <em>Home Care for the Dying Child: Professional and Family Perspectives.</em></td>
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<td>1983</td>
<td>Medicare hospice benefit created.</td>
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<td>1983</td>
<td>Children’s Hospice International (CHI) founded as a nonprofit organization to provide resources and support to children with life-threatening conditions and their families and to provide education, training, and technical assistance to healthcare professionals.</td>
</tr>
<tr>
<td>1983</td>
<td>Elizabeth Kubler-Ross publishes <em>On Children and Death.</em></td>
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<td>1984</td>
<td>The International Hospice Institute was founded, which later became the International Association for Hospice and Palliative Care.</td>
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<tr>
<td>1985</td>
<td>St. Mary’s Hospital for Children in New York initiates a comprehensive inpatient palliative care program.</td>
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<tr>
<td>1985</td>
<td>The Hospice Education Institute is founded to inform, educate, and support those seeking or providing care for dying people and the bereaved, or themselves coping with advanced illness or loss.</td>
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<tr>
<td>1988</td>
<td>The Academy of Hospice Physicians is founded and becomes the American Academy of Hospice and Palliative Medicine in 1996 (AAHPM).</td>
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<td>1988</td>
<td>The Association for Children’s Palliative Care is founded in England.</td>
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<td>1990</td>
<td>The World Health Organization (WHO) defines palliative care.</td>
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<td>1993</td>
<td>Ann Armstrong-Dailey and Sarah Zarbock from Children’s Hospice International publish <em>Hospice Care for Children</em>; the 2nd edition is published in 2001.*</td>
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<td>1995</td>
<td>The National Hospice and Palliative Care Organization (NHPCO) develops the first guidelines for palliative care eligibility for noncancer diagnoses.</td>
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<td>1995</td>
<td>Canuck Place Children’s Hospice opens in Vancouver, Canada.</td>
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<td>1995</td>
<td>CHI develops the Program for All-inclusive Care for Children and Their Families (PACC) to explore creative approaches to addressing the comprehensive needs of children with life-threatening conditions and their families from the time of diagnosis throughout the entire continuum of care in a cost-effective manner, maintaining the hope of cure and providing extensive bereavement support if cure is not attainable.</td>
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<tr>
<td>1997</td>
<td>The American Association of Colleges of Nursing developed recommended competencies and guidelines for nursing education and end-of-life care, <em>Peaceful Death: Recommended Competencies and Curricular Guidelines for End-of-Life Nursing Care,</em> which is revised in 2004.*</td>
</tr>
<tr>
<td>1998</td>
<td>Children’s Project on Palliative/Hospice Services is founded to identify critical issues in the field of pediatric palliative care and develop strategies to address these issues.</td>
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continued
Table 13-1. Historical Milestones in the Evolution of Pediatric Palliative Care

<table>
<thead>
<tr>
<th>Year</th>
<th>Milestone</th>
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<tbody>
<tr>
<td>2000</td>
<td>City of Hope and the American Association of Colleges of Nursing develop End of Life Nursing Education Curriculum (ELNEC) courses.</td>
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<td>2001</td>
<td>The Pediatric Hospice Palliative Care Special Interest Group of the Canadian Hospice Palliative Care Association defines pediatric palliative care; this group later becomes The Canadian Network of Palliative Care for Children.</td>
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<tr>
<td>2001</td>
<td>The Association for Children with Life-Threatening or Terminal Conditions and Their Families (ACT), the National Council for Hospice and Specialist Palliative Care Services, and the Scottish Partnership Agency for Palliative Care publishes Palliative Care for Young People Aged 13-24 years.</td>
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<td>2002</td>
<td>WHO amends the general definition of palliative care and adds issues specific to children.</td>
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<tr>
<td>2002</td>
<td>The first ELNEC pediatric-specific training course is developed.</td>
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<td>2002</td>
<td>AAHPM, the Hospice and Palliative Care Nurses Association, and NHPCO form the Hospice and Palliative Care Coalition.</td>
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<td>2002</td>
<td>The International Society for Paediatric Oncology adopts Guidelines for Assistance to Terminally Ill Children with Cancer, which recognizes the importance of symptom control and palliative care in pediatric oncology.</td>
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<td>2003</td>
<td>Last Acts, a national coalition to improve care near the end of life, publishes Precepts of Palliative Care for Children, Adolescents and Their Families developed by the Association of Pediatric Oncology Nurses, the Society of Pediatric Nurses, and the National Association of Neonatal Nurses.</td>
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<td>2004</td>
<td>The George Mark Children’s House in Northern California opens, the first and only free-standing residential pediatric palliative care center in the United States.</td>
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<td>2006</td>
<td>Launch of the “Partnership for Parents” website (<a href="http://www.partnershipforparents.org">www.partnershipforparents.org</a>) by the Children’s Hospice and Palliative Care Coalition (CHPCC) to offer support for parents whose children have a serious illness and for families grieving the loss of a child.</td>
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<tr>
<td>2006</td>
<td>Palliative care is recognized as a subspecialty by the American Board of Medical Specialties.</td>
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<tr>
<td>2008</td>
<td>Founding of the Canadian Network of Pediatric Hospices fosters collaboration among pediatric hospices in Canada.</td>
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<tr>
<td>2010</td>
<td>CureSearch Children’s Oncology Group and the Association of Pediatric Hematology/Oncology Nurses publish Pediatric Oncology Palliative and End-of-Life Care Resource.</td>
</tr>
<tr>
<td>2011</td>
<td>Joanne Wolfe, Pamela Hinds, and Barbara Sourkes publish Textbook of Interdisciplinary Pediatric Palliative Care with a Comprehensive Team Approach to the Unique Needs of Critically Ill Children.</td>
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PART IV
DISEASES
CASE STUDY

Chloe is a 9-year-old girl who was diagnosed with sickle cell disease at birth through the newborn screening program in her state. She has experienced several complications related to her sickle cell disease, including multiple admissions for vaso-occlusive crisis and for splenic sequestration and acute chest syndrome, both of which required blood transfusions. She presented to the emergency department (ED) with right lower-extremity pain and chest pain that had persisted for 2 days after she had played in the snow 3 days before. She had not experienced any fever, cough, or upper respiratory symptoms. Her mother treated Chloe’s pain with acetaminophen with codeine by mouth every 6 hours without any relief of the leg or chest pain.

Her physical examination revealed that Chloe walked with a limp in her right leg, and she reported leg pain as 9 on a scale of 1 to 10 and chest pain as 5 on a scale of 1 to 10. Her vital signs were stable, and her oxygen saturation was 98%. In the ED she was given morphine, 2 mg intravenously (IV), and ketorolac, 10 mg IV, for treatment of pain; she was started on hydration fluids of dextrose 5% in water + 0.45 normal saline at one and one-half times the maintenance dose. Her pain did not improve and was rated in the 8/10 to 10/10 range during her time in the ED. Complete blood count (CBC) results showed no significant decrease in her baseline hemoglobin level. Her urinalysis and electrolytes were within normal limits. While she was in the ED, she developed a fever of 102°F. A chest radiograph revealed left basilar airspace disease consistent with pneumonia/acute chest syndrome and stable mild cardiomegaly.

Chloe was admitted to the hematology/oncology unit, and treatment with azithromycin and ceftriaxone began. Blood cultures taken in the ED did not show any growth. During the hospital stay, she did not participate in recommended preventive care measures, such as using the incentive spirometer every 2 hours while she was awake and every 4 hours during the night or participating in physical therapy. She did use heat packs and attended whirlpool treatments to help relieve the pain. Unfortunately, both of Chloe’s parents had to work and were therefore unavailable to help support her during the hospitalization and encourage her to participate in care activities.

On the third hospital day, Chloe’s hemoglobin dropped to 6.0 g/dl and she developed respiratory distress. A second chest radiograph taken at that time revealed worsening infiltrate. She required 0.5 L of oxygen via nasal cannula to maintain her oxygen saturation at greater than 95%. She received a 10 ml/kg packed red blood cell transfusion, which increased her hemoglobin to 10 g/dl. By the next day her respiratory distress dissipated, and the oxygen was discontinued. She was able to maintain her oxygen saturation at greater than 95% on room air. Her pain was adequately treated with morphine delivered via patient-controlled analgesia (PCA), which allowed her to transition to oral oxycodone on hospital day 5. She developed severe constipation secondary to the opioids
and required an enema before discharge. She was discharged home on day 6 and returned to the clinic for follow-up a week later.

At her follow-up clinic appointment, the nurse practitioner reviewed possible triggers for vaso-occlusion, including exposure to extreme temperatures. She recommended that in the future, when Chloe wanted to play in the snow, she should dress warmly and come indoors frequently to allow her body to warm up. She should do this whenever she spends extended periods in the cold—whether she plays in the snow or swims in cold water. To prevent future complications, she should avoid activities in extreme weather or high altitudes, such as those encountered during hiking or skiing. The nurse practitioner also explained to Chloe the importance of her participation in her hospital care—by using the incentive spirometer and participating in physical therapy and other recommended activities to help her get better. She also talked with Chloe’s parents, acknowledging the difficulty of working while having a sick child in the hospital. She recommended that they identify another adult family member or friend who could help them by staying with Chloe for emotional support during hospitalizations.

So far, Chloe has not required any additional hospitalizations for vaso-occlusions or acute chest syndrome. Her team continues to follow her closely and is discussing the possibility of starting Chloe on hydroxyurea because of her repeated episodes of acute chest syndrome, which place her at risk of developing pulmonary complications. Chloe continues to attend school and participates in many activities but is careful to maintain good hydration whenever she anticipates exposure to extreme temperatures and allows herself rest periods during rigorous physical activity. Her parents were given a letter to submit to Chloe’s teachers, recommending that she be allowed to drink extra fluids, have additional bathroom time, and take frequent rest periods to prevent complications from her chronic disease. Chloe plans to attend a sickle cell camp next summer, which will give her an opportunity to interact with children like herself, learn more about sickle cell disease, and help foster her self-esteem and self-reliance as well as increase her participation in self-care activities.

INTRODUCTION
Nurses who care for pediatric patients with cancer and blood disorders frequently encounter red cell abnormalities in their patient population. In some instances, the primary concern of the patient involves a specific red cell disorder. However, some patients may also develop red cell disorders either because of a malignancy or as a result of treatment. It is imperative that nurses have a strong understanding of the etiology, symptoms, and treatments of pediatric red cell disorders to provide optimal care to their patients.

PHYSIOLOGY OF RED BLOOD CELLS
Hematopoiesis
Hematopoiesis begins in the embryonic yolk sac during the 3rd week of gestation. The liver and spleen become the predominant sites of hematopoiesis by the 12th week of gestation, and by the 30th week, the bone marrow predominates as the major site. Hematopoiesis starts with the pluripotent stem cell, which differentiates into either lymphoid or myeloid cells. The myeloid progenitor gives rise to erythrocytes as well as megakaryocytes, monocytes, and granulocytes. The first morphologically identifiable erythroid cell is the erythroblast, which undergoes enucleation (ie, removal of the nucleus) to become a reticulocyte, which then matures into the erythrocyte, or red blood cell (RBC). Erythropoietin (EPO) is a glycoprotein hormone that is necessary for the survival, proliferation, and differentiation of progenitors of the erythroid lineage. Thus, this hormone has been recognized as the physiologic regulator of red cell production. Its primary functions are to maintain a constant RBC mass and hemoglobin level as well as to hasten recovery of RBC loss after hemorrhage. EPO is produced in the kidney in adults and in the fetal liver in response to hypoxia and binds to erythropoietin receptors on red blood cells. EPO expression is activated when the partial pressure of oxygen (PO2) declines or when oxygen affinity increases. Its action is augmented by the hormones testosterone, somatotropin, and insulin-like growth factor 1. Erythropoiesis is stimulated by androgens (ie, male hormones) and inhibited by estrogens (ie, female hormones). For this reason, hemoglobin concentrations and RBC counts typically are greater in males than they are in females.
The RBC is the predominant cellular element in blood and has an average life span of 120 days. It delivers and releases oxygen to the tissues to meet their metabolic demands. The major component of the RBC is hemoglobin, an iron-containing protein that binds oxygen in the lungs and transports it to the tissues. Hemoglobin is comprised of an iron-containing heme ring and four globin (protein) chains, which comprise the protein component within the cell. Figure 21-1 depicts the structure of a hemoglobin molecule.

During fetal development, the predominant hemoglobin is fetal hemoglobin (HbF), which is composed of two α globin chains and two γ globin chains. HbF is remarkably resistant to denaturation at extreme pH levels, and it has a greater affinity for oxygen than maternal (adult) RBCs. The greater affinity for oxygen allows the fetus to more readily bind oxygen from maternal circulation. As early as the 14th week of gestation, β globin chains are activated, giving rise to the primary postnatal hemoglobin, HbA, which contains two α globin chains and two β globin chains. A switch from γ chain synthesis to β chain synthesis occurs just before birth in healthy individuals without hemoglobinopathies. The amount of HbF in neonates ranges from 65% to 95% and decreases steadily during the first year of life.

Therefore, the RBCs of newborns contain HbF in addition to small amounts of HbA and even smaller amounts of HbA₂, which is functionally similar to HbA. The switch from predominantly HbF to predominantly HbA is mostly complete by the age of 6 months in healthy newborns. This switch is well illustrated in Figure 21-2.

Anemia. Anemia is defined as reduction in red cell mass or hemoglobin concentration. It is the most common pediatric hematologic abnormality. Signs and symptoms of anemia include, but are not limited to, fatigue, pallor, tachycardia, headache, and shortness of breath. In addition to assessment for these signs and symptoms, a careful and detailed history can often identify potential causes (refer to Table 21-1). A number of potential causes are considered when gathering initial differential diagnoses, including congenital, acquired, benign, malignant, common, and rare disorders.

The initial steps in diagnosing anemia include a careful evaluation of the complete blood count (CBC), RBC indices, reticulocyte count, and peripheral blood smear. Because hemoglobin and hematocrit values and the red cell indices vary not only with age (refer to Table 21-2) but also among different laboratories, anemia is generally defined as a hemoglobin level that is

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**Figure 21-1. Structure of a Hemoglobin Molecule**

![Image of Hemoglobin Molecule]

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two standard deviations below the mean for the normal population. Different causes and types of anemia are presented throughout this chapter and are grouped according to the distinguishing characteristics they have in common. The section that follows describes in detail the different red cell indices that are evaluated when determining the cause of anemia.

Red blood cell indices. The RBC indices are groups of tests that provide information about the size, shape, and hemoglobin content of RBCs. These measures help the clinician to determine the cause of anemia and other red cell disorders by narrowing potential causes based on the red cell indices characteristic of specific types of anemia. The red cell indices include the mean cell volume (MCV), the mean corpuscular hemoglobin concentration (MCHC), and the red cell distribution width (RDW). The MCV is the measure of red cell volume and is the most useful of the red cell indices. The MCV is used to classify anemia as microcytic, low MCV—smaller than normal cells; normocytic, normal MCV—normal-sized cells; or macrocytic, elevated MCV—larger than normal cells.

In children younger than 10 years, the lower normal limit of the MCV is roughly 70 mcm³ plus the child’s age in years. The upper normal limit is 84 mcm³ + 0.6 mcm³ per year after 6 months of age until the adult upper normal limit of 96 mcm³ is reached. The mean corpuscular hemoglobin (MCH) and MCHC are generally less helpful in the diagnostic process. The MCH usually parallels the MCV, whereas the MCHC is an indicator of cellular hydration. The RDW is an indication of the variation in size of RBCs. The term used for a wide variation in RBC size is anisocytosis. Normal RDW values can vary depending on the laboratory equipment and indices used. The validity of different hemoglobin and RDW indices were compared in a group of 415 participants. This total number of participants consisted of four groups, including a control group, a group of patients with β-thalassemia trait, a group with α-thalassemia trait, and a group with iron deficiency anemia (IDA). The three indices used were the RBC distribution width standard deviation (RDW-SD), the RBC distribution width coefficient variation, and hemoglobin distribution width. Lin and colleagues concluded that

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Figure 21-2. Changing Patterns of Hemoglobin From Conception to 42 Weeks of Age

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