CHAPTER 34 Nursing Care of Clients with Hematologic Disorders

LEARNING OUTCOMES

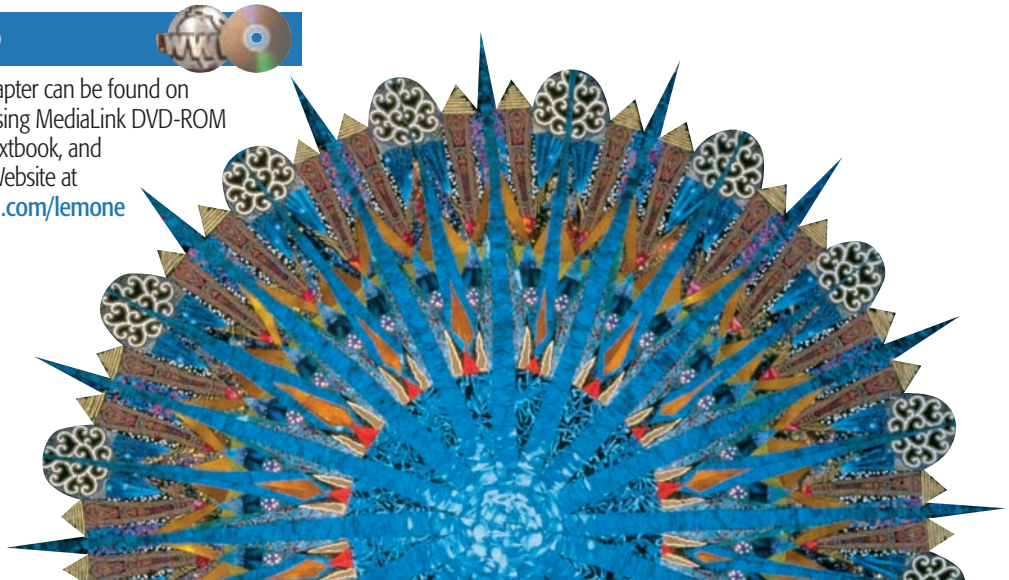
- Relate the physiology and assessment of the hematologic system and related systems to commonly occurring hematologic disorders.
- Describe the pathophysiology of common hematologic disorders.
- Explain nursing implications for medications and other treatments prescribed for clients with hematologic disorders.
- Discuss indications for and complications of bone marrow or stem cell transplantation, as well as related nursing care.
- Compare and contrast the pathophysiology, manifestations, and management of bleeding disorders.
- Describe the major types of leukemia and the most common treatment modalities and nursing interventions.
- Differentiate Hodgkin's disease from non-Hodgkin's lymphomas.

CLINICAL COMPETENCIES

- Assess effects of hematologic disorders and prescribed treatments on clients' functional health status.
- Monitor and document continuing assessment data, including laboratory test results, subjective and objective information, reporting data outside the normal or expected range.
- Based on knowledge of pathophysiology, prescribed treatment, and assessed data, identify and prioritize nursing diagnoses for clients with hematologic disorders.
- Use nursing research and evidence-based practice to identify and implement individualized nursing interventions for the client with a hematologic disorder.
- Safely and knowledgeably administer prescribed medications and treatments for clients with hematologic disorders.
- Collaborate with the interdisciplinary care team to plan and provide coordinated, effective care for clients with hematologic disorders.
- Provide appropriate teaching for clients with hematologic disorders, evaluating learning and the need for continued reinforcement of information.
- Use continuing assessment data to revise the plan of care as needed to restore, maintain, or promote functional health in the client with a hematologic disorder.

MEDIALINK

Resources for this chapter can be found on the Prentice Hall Nursing MediaLink DVD-ROM accompanying this textbook, and on the Companion Website at http://www.prenhall.com/lemone



KEY TERMS

anemia, *1102*
aplastic anemia, *1109*
bone marrow transplant (BMT), *1124*
disseminated intravascular coagulation (DIC), *1146*
hemolytic anemias, *1106*
hemophilia, *1142*
hemostasis, *1139*
iron deficiency anemia, *1103*
leukemia, *1118*
lymphoma, *1129*
multiple myeloma, *1136*
pernicious anemia, *1105*
polycythemia, *1117*
sickle cell anemia, *1106*
sickle cell crisis, *1107*
stem cell transplant (SCT), *1124*
thalassemia, *1109*
thrombocytopenia, *1139*

Disorders affecting the blood and blood-forming organs have effects that range from minor disruptions in daily activities to major life-threatening crises. Clients with hematologic disorders need holistic nursing care, including emotional support and care for problems involving major body systems.

This chapter focuses on health changes resulting from changes in red cells, white cells, platelets, and clotting factors. Before proceeding with this chapter, read Chapter 33 ∞, which provides a review of the physiology of blood and its formation, as well as important information about assessing clients with hematologic disorders.

RED BLOOD CELL DISORDERS

Red blood cells (RBCs) transport oxygen to body tissues and help return carbon dioxide to the lungs for excretion. Alterations in the number, size, shape, or composition of RBCs affect their ability to effectively carry out these functions. Anemia, the most common RBC disorder, is an abnormally low RBC count or reduced hemoglobin content. Polycythemia is an abnormally high RBC count.

THE CLIENT WITH ANEMIA

Anemia is an abnormally low number of circulating RBCs, low hemoglobin concentration, or both. Decreased numbers of circulating RBCs is the usual cause of anemia. This may result from blood loss, inadequate RBC production, or increased RBC destruction. Insufficient or defective hemoglobin within RBCs contributes to anemia. Depending on its severity, anemia may affect all major organ systems.

FAST FACTS

- Iron deficiency anemia, a nutritional anemia, is the most common type of anemia.
- Blood loss anemia may be either acute, resulting from hemorrhage, or chronic, resulting from chronic blood loss (e.g., menstrual flow, slow GI bleeding).

Physiology Review

As blood flows through the pulmonary vascular system, oxygen diffuses from alveoli into capillary blood. The majority of the oxygen binds reversibly with the hemoglobin in red blood cells; only about 3% of the oxygen remains in solution in the blood. When the blood reaches the capillaries serving body tissues, oxygen is released from the hemoglobin molecule, and diffuses out of the capillary to reach the cells. The amount of oxygen that reaches the tissues depends on a number of factors, including:

- Available oxygen in the alveoli
- The diffusing surface and capacity of the lungs
- The number of red blood cells and the amount and type of hemoglobin they contain
- The ability of the cardiovascular system to transport blood and oxygen to the tissues.

For more information about red blood cells, hemoglobin, and their production and function, see Chapter 33 ∞.

Physiology and Manifestations

A number of different pathologic mechanisms can lead to anemia (Box 34–1). Regardless of the cause, every type of anemia reduces the oxygen-carrying capacity of the blood due to a deficiency of RBCs or hemoglobin, leading to tissue hypoxia. The resulting manifestations depend on the severity of the anemia, how quickly it develops, and other factors such as age and health status.

When anemia develops gradually and the RBC reduction is moderate, successful compensatory mechanisms may result in few symptoms except when the oxygen needs of the body increase due to exercise or infection. Symptoms develop as RBCs and hemoglobin levels are further reduced. Pallor of the skin, mucous membranes, conjunctiva, and nail beds develops as a result of blood redistribution to vital organs and lack of hemoglobin (Figure 34–1 ■). As tissue oxygenation decreases, the heart and respiratory rates rise in an attempt to increase cardiac output and tissue perfusion. Tissue hypoxia may cause angina, fatigue, dyspnea on exertion, and night cramps. It also stimulates erythropoietin release; increased erythropoietin activity stimulates RBC production in the bone marrow, and may lead to bone pain. Cerebral hypoxia can lead to headache, dizziness, and dim vision. Heart failure may develop in severe anemia.

With rapid blood loss, blood volume is decreased as well as the oxygen-carrying capacity of the blood. Initial manifestations include tachycardia and tachypnea; the skin may be pale, cool, and clammy as peripheral vessels constrict to maintain blood flow to the heart and brain. With significant blood loss, signs of circulatory shock may occur, including hypotension,

BOX 34–1 Pathophysiologic Mechanisms of Anemia

Decreased RBC Production

- Altered hemoglobin synthesis
  - Iron deficiency
  - Thalassemias
  - Chronic inflammation
- Altered DNA synthesis
  - Vitamin B_{12} or folic acid malabsorption or deficiency
- Bone marrow failure
  - Aplastic anemia (stem cell dysfunction)
  - Red cell aplasia
  - Myeloproliferative leukemias
  - Cancer metastasis, lymphoma
  - Chronic infection or inflammation, physical and emotional fatigue

Increased RBC Loss or Destruction

- Acute or chronic blood loss
  - Hemorrhage or trauma
  - Chronic gastrointestinal bleeding, menorrhagia
- Increased hemolysis
  - Hereditary cell membrane disorders
  - Defective hemoglobin—sickle cell anemia or trait
  - Pyruvate kinase (PK) or G6PD deficiency affecting glycolysis or cell oxidation
  - Immune mechanisms and disorders (e.g., blood reaction, hypersensitivity responses, autoimmune disorders)
  - Splenomegaly and hypersplenism
  - Infection
  - Erythrocyte trauma (e.g., due to cardiopulmonary bypass, hemolytic uremic syndrome)

tachycardia, decreased level of consciousness, and oliguria. With chronic bleeding, fluid shifts from the interstitial spaces into the vessels, maintaining blood volume. Blood viscosity is reduced, which may result in a systolic heart murmur. See page 1104 for *Multisystem Effects of Anemia.*

Anemia is categorized by cause: blood loss, nutritional, hemolytic, and bone marrow suppression. The pathophysiology and specific manifestations of these types of anemias follow.

Blood Loss Anemia

When anemia results from acute or chronic bleeding, RBCs and other blood components (such as iron) are lost from the body. With acute blood loss, circulating volume decreases. As a result, the cardiac output falls. Compensatory mechanisms are activated to maintain the cardiac output: the heart rate increases, and peripheral blood vessels constrict. Vessels in the liver, a blood storage organ, also constrict, increasing circulating volume. Fluid shifts from the interstitial spaces into the vascular compartment to maintain blood volume, diluting the cellular components of the blood and reducing its viscosity. If hemorrhage continues, compensatory mechanisms become less effective, increasing the risk for shock and circulatory failure (see Chapter 11 ∞).

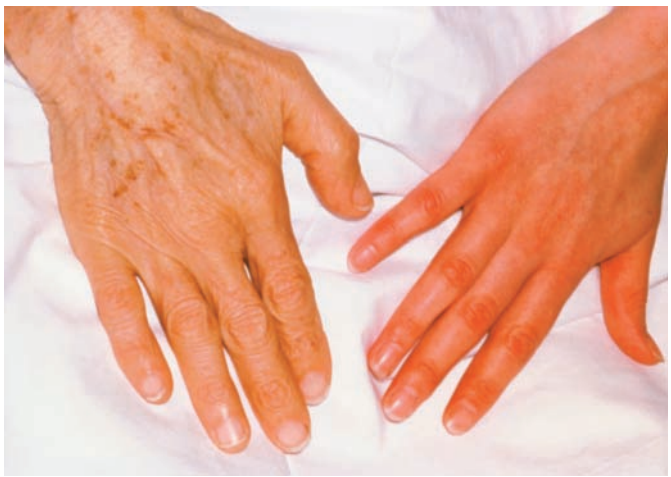


Figure 34–1 ■ The skin of the client with anemia appears pale beside that of a person with a normal hemoglobin and hematocrit.

Source: Westminster Hospital, Photo Researchers, Inc.

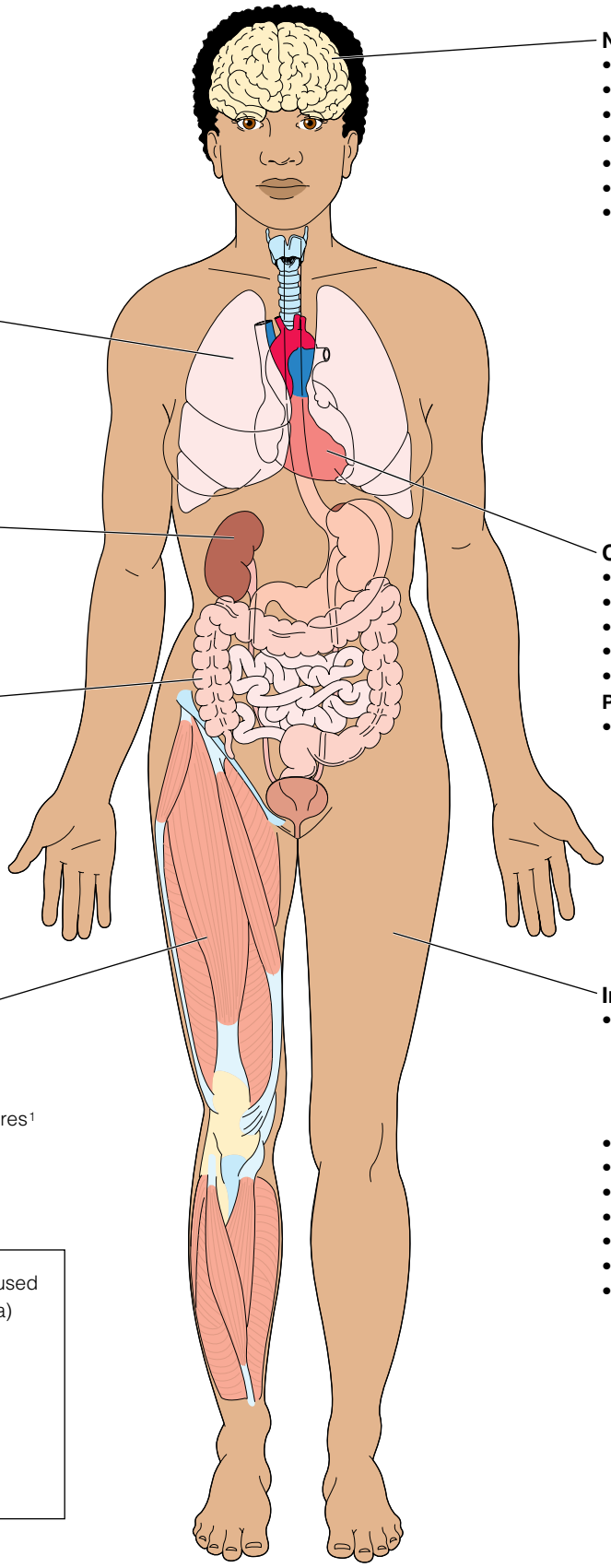
In acute blood loss, circulating RBCs are of normal size and shape (*normocytic*). Early in the hemorrhage, the RBC count, hemoglobin, and hematocrit may be normal; as fluid shifts from the interstitial space into the vascular space to maintain circulating volume, the RBC count, hemoglobin, and hematocrit fall. If sufficient iron is available, the number of circulating RBCs and hemoglobin levels return to normal within 3 to 4 weeks after the bleeding episode. Chronic blood loss, on the other hand, depletes iron stores as RBC production attempts to maintain the RBC supply. The resulting RBCs are *microcytic* (small) and *hypochromic* (pale).

Nutritional Anemias

A number of different nutrients are required for normal red blood cell development (erythropoiesis). Iron is a key nutrient necessary for hemoglobin synthesis. In addition, adequate supplies of protein (and its building blocks, amino acids), vitamins, and other minerals are required. The B vitamins, particularly B_{12} (cobalamin) and folate, play a key role in RBC development. Vitamins C and E also are necessary. Nutritional anemias result from nutrient deficits that affect RBC formation or hemoglobin synthesis. The nutrient deficit may be caused by inadequate diet, malabsorption of the nutrient, or an increased need for the nutrient. The most common types of nutritional anemias are iron deficiency anemia, vitamin B_{12} anemia, and folic acid deficiency anemia. Vitamin B_{12} and folic acid anemias are sometimes called *megaloblastic* anemias, because enlarged nucleated RBCs called megaloblasts are seen in these anemias.

IRON DEFICIENCY ANEMIA **Iron deficiency anemia** is the most common type of anemia. It develops when the supply of iron is inadequate for optimal RBC formation. The body cannot synthesize hemoglobin without iron. Normally, the body efficiently recycles and stores iron, reusing much of the iron contained in RBCs that are removed from circulation due to age or damage. However, small amounts of iron continually are lost

MULTISYSTEM EFFECTS **of Anemia**



Neurologic
- Paresthesias [4]
- Proprioception deficits [4]
- Headache [5]
- Fainting [5]
- Forgetfulness [5]
- Pain [6]
- Behavioral disturbances (pica) [3]

Respiratory
- Increased rate
- Dyspnea on exertion

Urinary
- Hemoglobinuria [7]

Gastrointestinal
- Diarrhea [4]
- Anorexia [5]
- Nausea [5]
- Gallstones [6]
- Splenomegaly [1]
- Abdominal pain [6]

Cardiovascular
- Tachycardia
- Palpitations
- Systolic murmur
- Ventricular hypertrophy
- Angina

Potential complication
- Heart failure [6]

Musculoskeletal
- Night cramps [5]
- Bone pain
- Joint pain [6]
- Bone deformity and fractures [1]

Integumentary
- Pallor
  - Skin
  - Mucous membranes
  - Conjunctiva
  - Nail beds
- Jaundice [1]
- Petechiae [2]
- Purpura [2]
- Spoon-shaped nails [3]
- Cheilosis [3]
- Sore, beefy red tongue [4]
- Chronic leg ulcers [6]

Key (symptoms usually caused by a specific form of anemia)
1 Hemolytic anemias
2 Aplastic anemia
3 Iron deficiency anemia
4 Pernicious anemia
5 Vitamin B_{12} anemia
6 Sickle cell anemia
7 G6PD anemia

in the feces; therefore, adequate iron intake is necessary for normal hemoglobin synthesis and RBC production. Iron deficiency anemia results in fewer numbers of RBCs, microcytic and hypochromic RBCs, as well as malformed RBCs (*poikilocytosis*) (Figure 34–2 ■).

Excessive iron loss due to chronic bleeding is the usual cause of iron deficiency anemia in adults. Menstrual blood loss is the most common cause in adult females. Iron deficiency anemia also may result from inadequate dietary iron intake (less than 1 mg/day), malabsorption syndromes, or the increased iron requirements associated with pregnancy and lactation. Box 34–2 summarizes common causes of iron deficiency anemia.

Iron deficiency anemia is particularly common in older adults. Chronic, occult (hidden) blood loss may occur from slowly bleeding peptic ulcers, gastrointestinal inflammation, hemorrhoids, and cancer. Inadequate dietary iron intake also contributes to anemia in the older adult. Access to transportation may limit fresh food consumption, a factor contributing to poor iron intake among all adults, especially people with limited or fixed incomes.

Manifestations In addition to the general manifestations of anemia described earlier, chronic iron deficiency may lead to brittle, spoon-shaped nails; cheilosis (cracks at the corners of the mouth); a smooth, sore tongue; and *pica* (a craving for unusual substances, such as clay or starch).

VITAMIN B_{12} DEFICIENCY ANEMIA Vitamin B_{12} is necessary for DNA synthesis and is almost exclusively found in foods derived from animals. Vitamin B_{12} deficiency occurs when inadequate vitamin B_{12} is consumed or, more commonly, when it is poorly absorbed from the gastrointestinal tract. Deficiency of this vitamin impairs cell division and maturation of the cell nucleus, especially in rapidly proliferating red blood cells. As a result, macrocytic (large), misshapen (oval rather than concave) RBCs with thin membranes are produced. Great numbers of these large, immature RBCs enter the circulation. These cells are fragile, incapable of carrying adequate amounts of oxygen, and have a shortened life span.

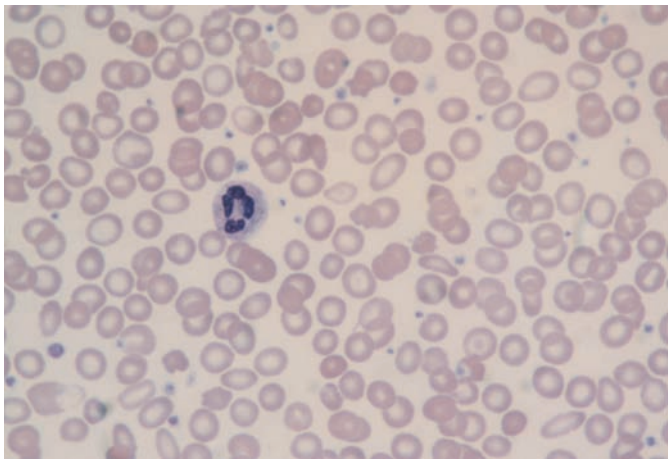


Figure 34–2 ■ A blood smear showing RBCs characteristically seen in iron deficiency anemia. Note the pale color of the RBCs (hypochromic). Many of the cells also are smaller than normal (microcytic) and misshapen, reducing their oxygen-carrying capacity.

Source: Dr. E. Walker, Photo Researchers, Inc.

BOX 34–2 Causes of Iron Deficiency Anemia

- Dietary deficiencies
  - Vegetarian diet
  - Inadequate protein intake
- Decreased absorption
  - Partial or total gastrectomy
  - Chronic diarrhea
  - Malabsorption syndromes
- Increased metabolic requirements
  - Pregnancy
  - Lactation
- Blood loss
  - Gastrointestinal bleeding (especially due to ulcers or chronic aspirin use)
  - Menstrual losses
- Chronic hemoglobinuria

Failure to absorb dietary vitamin B_{12} is called **pernicious anemia**. It develops due to lack of *intrinsic factor,* a substance secreted by the gastric mucosa. Intrinsic factor binds with vitamin B_{12} and travels with it to the ileum, where the vitamin is absorbed. In the absence of intrinsic factor, vitamin B_{12} cannot be absorbed into the body.

Vitamin B_{12} deficiency may also result from other malabsorption disorders and dietary factors. Resection of the stomach or ileum, loss of pancreatic secretions, and chronic gastritis can affect vitamin B_{12} absorption. Dietary deficiencies of vitamin B_{12} are rare, usually occurring only among strict vegetarians.

Manifestations Manifestations of vitamin B_{12} deficiency anemia develop gradually as bodily stores of the vitamin are depleted. Pallor or slight jaundice and weakness develop. In pernicious anemia, a smooth, sore, beefy red tongue and diarrhea may occur. Because vitamin B_{12} is important for neurologic function, *paresthesias* (altered sensations, such as numbness or tingling) in the extremities and problems with *proprioception* (the sense of one's position in space) develop. These manifestations may progress to difficulty maintaining balance due to spinal cord damage. Central nervous system (CNS) manifestations of relatively short duration (6 months or less) are reversible with treatment, but may be permanent if treatment is delayed (Tierney et al., 2005).

FOLIC ACID DEFICIENCY ANEMIA Like vitamin B_{12}, folic acid is required for DNA synthesis and normal maturation of red blood cells. Folic acid deficiency anemia is characterized by fragile, megaloblastic (large and immature) cells. Folic acid is found in green leafy vegetables, fruits, cereals, and meats, and is absorbed from the intestines.

Folic acid deficiency anemia due to inadequate intake is more common among people who are chronically undernourished. This includes older adults, alcoholics, and the drug addicted. Alcoholics are especially at risk because alcohol suppresses folate metabolism, which forms folic acid. Increased folic acid requirements also may lead to anemia. Pregnant women are at the

greatest risk. Infants and teenagers can develop temporary folic acid deficiencies during periods of rapid growth. Impaired folic acid absorption and metabolism can cause folic acid deficiency anemia. Malabsorption disorders such as celiac sprue (a hereditary gastrointestinal disorder characterized by inability to metabolize amino acids found in gluten), and certain medications, such as methotrexate and some chemotherapeutic agents, may be implicated. Causes of folic acid deficiency anemia are summarized in Box 34–3.

Manifestations The manifestations develop gradually as folic acid stores are depleted. Signs and symptoms may include pallor, progressive weakness and fatigue, shortness of breath, and heart palpitations. Manifestations similar to those associated with vitamin B_{12} anemia, such as glossitis, cheilosis, and diarrhea, are common. No neurologic symptoms occur with folic acid deficiency anemia, helping differentiate it from vitamin B_{12} deficiency anemia. These two nutritional anemias do, however, sometimes coexist.

Maternal folic acid deficiency is strongly associated with neural tube defects such as meningomyelocele. The neural tube develops early in the process of fetal development, often before pregnancy is recognized.

Hemolytic Anemias

Hemolytic anemias are characterized by premature destruction (*lysis*) of RBCs. When RBCs break down, iron and other by-products of their destruction remain in the plasma. RBC lysis (hemolysis) may occur within the circulatory system or due to phagocytosis by WBCs such as circulating monocytes and macrophages in the spleen. In response to hemolysis, the hematopoietic activity of bone marrow increases, leading to increased reticulocytes (immature RBCs) in circulating blood. Most types of hemolytic anemia are characterized by normocytic and normochromic RBCs.

There are many different causes of hemolytic anemias (Box 34–4). The cause may be *intrinsic,* arising from disorders within the RBC itself, or *extrinsic,* originating outside the RBC.

BOX 34–3 Causes of Folic Acid Deficiency Anemia

- Inadequate dietary intake
  - *At risk:*
  - a. Older adults
  - b. Alcoholics
  - c. Clients receiving total parenteral nutrition
- Increased metabolic requirements
  - *At risk:*
  - a. Pregnant women
  - b. Infants and teenagers
  - c. Clients undergoing hemodialysis
  - d. Clients with forms of hemolytic anemia
- Folic acid malabsorption and impaired metabolism
  - a. Celiac sprue
  - b. Chemotherapeutic agents, folate antagonists (methotrexate, pentamidine), or anticonvulsants
  - c. Alcoholism

BOX 34–4 Causes of Hemolytic Anemia

Intrinsic
- RBC cell-membrane defects
- Hemoglobin structure defects (e.g., sickle cell anemia, thalassemia)
- Inherited enzyme defects (e.g., G6PD deficiency)

Extrinsic
- Drugs, chemicals
- Toxins and venoms
- Bacterial and other infections
- Trauma, burns
- Mechanical damage (prosthetic heart valves)

Intrinsic disorders include cell membrane defects, defects in hemoglobin structure and function, and inherited enzyme deficiencies. See the accompanying Focus on Cultural Diversity box for more information about inherited intrinsic RBC disorders associated with hemolytic anemia. Extrinsic causes of hemolytic anemia include drugs, bacterial and other toxins, and trauma. This section discusses sickle cell anemia, thalassemia, acquired hemolytic anemia, and glucose-6-phosphate dehydrogenase anemia.

SICKLE CELL ANEMIA **Sickle cell anemia** is a hereditary, chronic hemolytic anemia. It is characterized by episodes of *sickling,* during which RBCs become abnormally crescent shaped. The disorder is transmitted as an autosomal recessive genetic defect (Figure 34–3 ■). This defect causes synthesis of an abnormal form of hemoglobin (HbS) within red blood cells. Sickle cell anemia can significantly shorten life span, with most deaths occurring due to infection (McCance & Huether, 2006).

The disease is most common among people of African descent (see Focus on Cultural Diversity box below). In the

FOCUS ON CULTURAL DIVERSITY Inherited Hemolytic Anemias

- Sickle cell anemia affects about 72,000 people in the United States
  - In African Americans, sickle cell anemia occurs in 1 out of every 600 births.
  - People from Central and South America, Cuba, Saudi Arabia, India, and Mediterranean countries such as Turkey, Greece, and Italy also may be at risk; sickle cell anemia occurs in 1 of every 1000 to 1400 Hispanic American births.
- Thalassemia is less common than sickle cell anemia.
  - Alpha-thalassemia primarily affects people of Southeast Asian, Indian, Chinese, or Filipino ancestry.
  - Beta-thalassemia is seen primarily in people of Mediterranean, Asian, and African origin.
- G6PD anemia primarily affects people of African or Mediterranean descent.
  - This hereditary defect is carried on the X chromosome and affects more men than women.

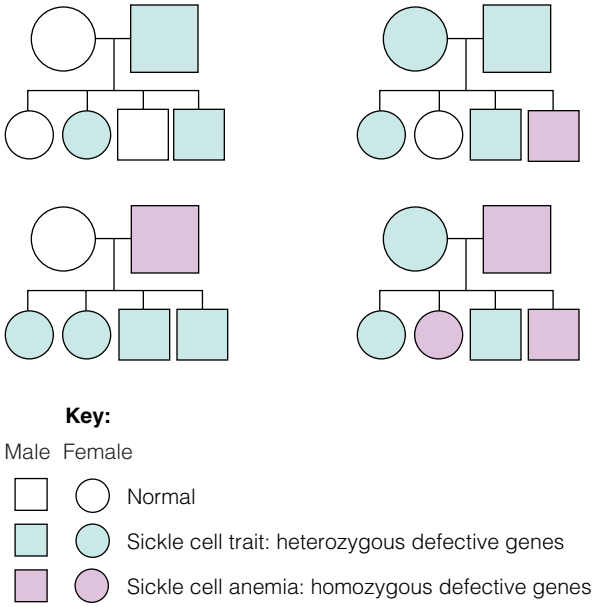


Figure 34–3 ■ Inheritance pattern for sickle cell anemia.

FOCUS ON CULTURAL DIVERSITY
Sickle Cell Anemia

Sickle cell anemia tends to affect people whose origins are in equatorial countries, particularly those in central Africa, the Near East, the Mediterranean region, and parts of India. Hispanics from the Caribbean and Central and South America also may have the HbS gene. This gene may protect against lethal forms of malaria, an endemic disease in many equatorial regions.

The gene for HbS is transmitted in an autosomal recessive pattern from parent to offspring. A parent with one HbS gene (heterozygous) has a 50% risk of transmitting the gene to each child (see Figure 34–3). If both parents carry the gene, each child has a 25% risk of inheriting the gene from both parents. A person who carries both HbS genes (homozygous) is likely to develop sickle cell disease.

Sickle cell anemia is a serious chronic and recurrent disease. The stress of the disease is compounded by the risk for its transmission to offspring. Recommend that all clients with sickle cell trait or disease obtain genetic counseling as part of their family planning process.

United States, 7% to 13% of blacks carry the defective gene, having inherited it from one parent (McCance & Huether, 2006). These people have *sickle cell trait.* About 40% of their hemoglobin is HbS (Porth, 2005). They are likely to remain asymptomatic unless stressed by severe hypoxia. Less than 1% of African Americans are homozygous for the disorder; that is, they have inherited a defective gene from both parents. These people have *sickle cell disease;* nearly all their hemoglobin is HbS (Porth, 2005). They are at risk for **sickle cell crisis**, severe episodes of fever and intense pain that are the hallmark of this disorder.

The HbS gene changes the structure of the beta chain of the hemoglobin molecule. When hypoxemia develops and HbS is deoxygenated, it crystallizes into rodlike structures. Clusters of these rods form long chains that deform the erythrocyte into a crescent or sickle shape (Figure 34–4 ■). The sickled cells tend to clump together and obstruct capillary blood flow, causing ischemia and possible infarction of surrounding tissue. See *Pathophysiology Illustrated: Sickle Cell Anemia* on page 1108.

When normal oxygen tension is restored, the sickled RBCs resume their normal shape; that is, they "unsickle." Repeated episodes of sickling and unsickling weaken RBC cell membranes. The weakened RBCs are hemolyzed and removed. Consequently, the normal life span of RBCs is greatly reduced in sickle cell anemia, increasing the demand for RBC production. Conditions likely to trigger sickling include hypoxia, low environmental or body temperature, excessive exercise, anesthesia, dehydration, infections, or acidosis.

Manifestations and Complications The acute and chronic manifestations of sickle cell anemia arise from episodes of RBC sickling. Sickling causes general manifestations of hemolytic anemia, including pallor, fatigue, jaundice, and irritability. Extensive sickling can precipitate a crisis due to occluded circulation, impaired erythropoiesis, or sequestration of large amounts of blood in the liver or spleen.

A vasoocclusive or thrombotic crisis occurs when sickling develops in the microcirculation. Obstruction of blood flow triggers vasospasm that halts all blood flow in the vessel. Lack of blood flow leads to tissue ischemia and infarction. Vasoocclusive crises are painful and last an average of 4 to 6 days. Infarction of small vessels in the extremities causes painful swelling of the hands and feet; large joints also may be affected. Priapism (persistent, painful erection of the penis) may develop. Abdominal pain may signal infarction of abdominal organs and structures. Infarction may affect bone marrow or lead to aseptic necrosis of affected bones. Stroke may result from cerebral vessel occlusion (McCance & Huether, 2006). Skin ulcers may develop as the result of

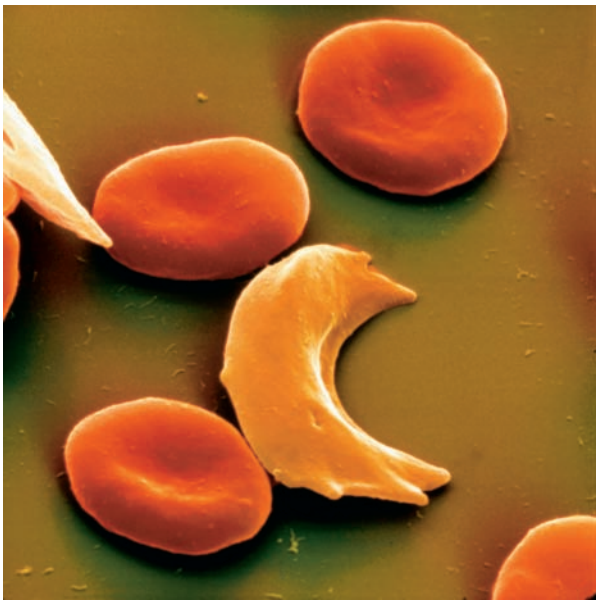


Figure 34–4 ■ Blood smear containing normal red blood cells and sickle cells.

Source: Oliver Meckes and Nicole Ottawa, Photo Researchers, Inc.

MEDIALINK Sickle Cell Anemia Animation

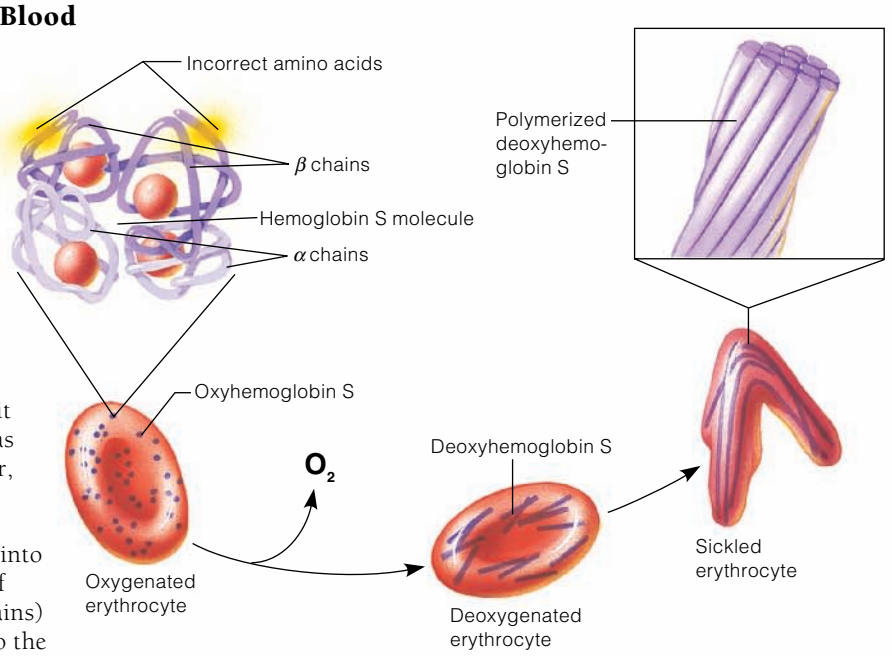
PATHOPHYSIOLOGY ILLUSTRATED

Sickle Cell Anemia

Hemoglobin S and Red Blood Cell Sickling

Sickle cell anemia is caused by an inherited autosomal recessive defect in Hb synthesis. Sickle cell hemoglobin (HbS) differs from normal hemoglobin only in the substitution of the amino acid valine for glutamine in both beta chains of the hemoglobin molecule.

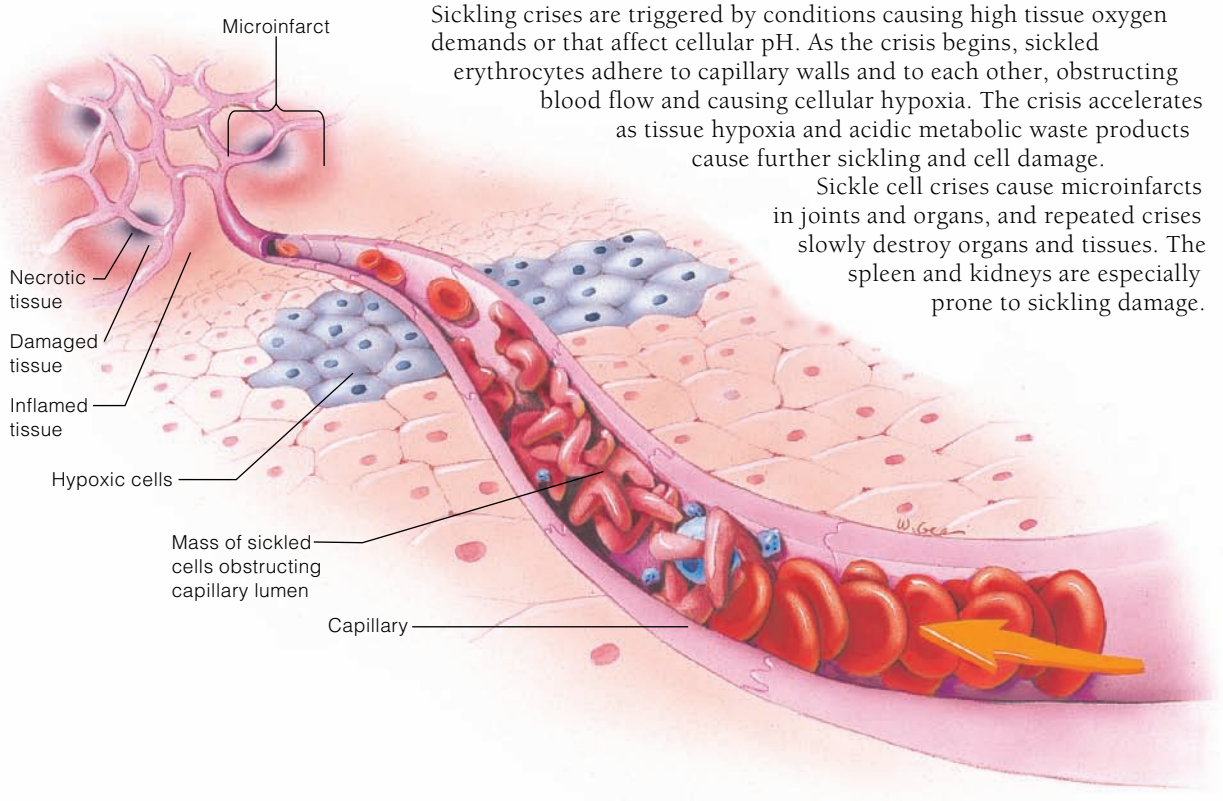
When HbS is oxygenated, it has the same globular shape as normal hemoglobin. However, when HbS off-loads oxygen, it becomes insoluble in intracellular fluid and crystallizes into rodlike structures. Clusters of rods form polymers (long chains) that bend the erythrocyte into the characteristic crescent shape of the sickle cell.



The Sickle Cell Disease Process

Sickle cell disease is characterized by episodes of acute painful crises. Sickling crises are triggered by conditions causing high tissue oxygen demands or that affect cellular pH. As the crisis begins, sickled erythrocytes adhere to capillary walls and to each other, obstructing blood flow and causing cellular hypoxia. The crisis accelerates as tissue hypoxia and acidic metabolic waste products cause further sickling and cell damage.

Sickle cell crises cause microinfarcts in joints and organs, and repeated crises slowly destroy organs and tissues. The spleen and kidneys are especially prone to sickling damage.



occluded vessels supplying the dermis. Repeated infarcts associated with sickling can affect the structure and function of nearly every organ system. People with sickle cell disease may develop an enlarged spleen and liver, renal insufficiency, gallstones, and other manifestations of organ dysfunction. *Acute chest syndrome*, a symptom complex that includes fever, chest pain, an increasing WBC count, and pulmonary infiltrates, may develop, as well as other pulmonary complications such as pneumonia, pulmonary infarction, and pulmonary embolism (Porth, 2005).

The shortened RBC life span and compromised erythropoiesis can lead to profound *aplastic anemia* in sickle cell disease. *Sequestration crises* are marked by pooling of large amounts of blood in the liver and spleen. This sickle cell crisis only occurs in children, but is thought to be the cause of sickle cell disease-related deaths in early childhood (Porth, 2005).

THALASSEMIA The **thalassemias** are inherited disorders of hemoglobin synthesis in which either the alpha or beta chains of the hemoglobin molecule are missing or defective. This leads to deficient hemoglobin production and fragile hypochromic, microcytic RBCs called *target cells* because of their distinctive bull's-eye appearance.

Thalassemia usually affects certain populations. People of Mediterranean descent (southern Italy and Greece) are more likely to have beta-defect thalassemias (often called *Cooley's anemia* or Mediterranean anemia). People of Asian ancestry, especially from Thailand, the Philippines, and China, more often have alpha-defect thalassemia. Africans and African Americans may have both alpha- and beta-defect thalassemia. As with sickle cell anemia, only one defective beta chain-forming gene may be present (*beta-thalassemia minor*), causing mild symptoms, or both may be defective (*beta-thalassemia major*), leading to more severe symptoms. Children with thalassemia major rarely reach adulthood, although repeated blood transfusions may extend their life span (McCance & Huether, 2006). Four genes are responsible for alpha chain formation; one, two, three, or all four may be defective. In the latter case (*alpha-thalassemia major*), death is inevitable and usually occurs *in utero.* Genetic studies and counseling are recommended for people at risk for this illness.

Manifestations and Complications People with thalassemia minor often are asymptomatic. When manifestations do occur, they include mild to moderate anemia, mild splenomegaly, bronze skin coloring, and bone marrow hyperplasia. The major form of the disease causes severe anemia, heart failure, and liver and spleen enlargement from increased red cell destruction. Fractures of the long bones, ribs, and vertebrae may result from bone marrow expansion and thinning due to increased hematopoiesis. Jaundice may develop due to hemolysis, as well as hepatomegaly and splenomegaly. Accumulation of iron in the heart, liver, and pancreas following repeated transfusions for treatment may eventually cause failure of these organs.

ACQUIRED HEMOLYTIC ANEMIA *Acquired hemolytic anemia* results from hemolysis due to factors outside of the RBC. Causes of acquired hemolytic anemias include:

- Mechanical trauma to RBCs produced by prosthetic heart valves, severe burns, hemodialysis, or radiation
- Autoimmune disorders
- Bacterial or protozoal infection
- Immune-system-mediated responses, such as transfusion reactions
- Drugs, toxins, chemical agents, or venoms.

The manifestations of acquired hemolytic anemia depend on the extent of hemolysis and the body's ability to replace destroyed RBCs. The anemia itself often is mild to moderate as erythropoiesis increases to replace the destroyed RBCs. The spleen enlarges as it removes damaged or destroyed RBCs. If the breakdown of heme units exceeds the liver's ability to conjugate and excrete bilirubin, jaundice develops. When the condition is severe, bone marrow expands, and bones may be deformed or may develop pathologic fractures. The severity of generalized manifestations of anemia (tachycardia, pallor, etc.) depends on the degree of anemia and deficiency of tissue oxygenation.

GLUCOSE-6-PHOSPHATE DEHYDROGENASE (G6PD) ANEMIA *Glucose-6-phosphate dehydrogenase (G6PD) anemia* is caused by a hereditary defect in RBC metabolism. It is relatively common in people of African and Mediterranean descent. The defective gene is located on the X chromosome and therefore affects more males than females. There are many variations of this genetic defect.

G6PD is an enzyme that catalyzes glycolysis, the process in which an RBC derives cellular energy. A defect in G6PD action causes direct oxidation of hemoglobin, damaging the RBC. Hemolysis usually occurs only when the affected person is exposed to stressors (e.g., drugs such as aspirin, sulfonamides, or vitamin K derivatives) that increase the metabolic demands on RBCs. The G6PD deficiency impairs the necessary compensatory increase in glucose metabolism and causes cellular damage. Damaged RBCs are destroyed over a period of 7 to 12 days.

When exposed to a stressor triggering G6PD anemia, symptoms develop within several days. These may include pallor, jaundice, hemoglobinuria (hemoglobin in the urine), and an elevated reticulocyte count. As new RBCs develop, counts return to normal.

Aplastic Anemia

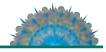
In **aplastic anemia**, the bone marrow fails to produce all three types of blood cells, leading to *pancytopenia.* Normal bone marrow is replaced by fat. Fortunately, aplastic anemia is rare. *Fanconi anemia* is a rare aplastic anemia caused by defects of DNA repair. The underlying cause of about 50% of acquired aplastic anemia is unknown (*idiopathic aplastic anemia*). Other cases follow stem cell damage caused by exposure to radiation or certain chemical substances such as benzene, arsenic, nitrogen mustard, certain antibiotics (especially chloramphenicol), and chemotherapeutic drugs (McCance & Huether, 2006). Aplastic anemia also may occur with viral infections such as mononucleosis, hepatitis C, and HIV disease (Porth, 2005).

In aplastic anemia, the number of stem cells in the bone marrow is significantly reduced. The stem cell pool may be less than 1% of normal when the disease is recognized. Anemia develops as the bone marrow fails to replace RBCs that have reached the end of their life span. Remaining RBCs may be

normochromic and normocytic or may be large with increased mean corpuscular volume.

Manifestations Manifestations of aplastic anemia vary with the severity of the pancytopenia. Its onset usually is insidious, but may be sudden. Manifestations include fatigue, pallor, progressive weakness, exertional dyspnea, headache, and ultimately tachycardia and heart failure. Platelet deficiency leads to bleeding problems; bleeding gums, excessive bruising, and nosebleeds may be the initial symptoms. A deficiency of white blood cells increases the risk of infection, causing manifestations such as sore throat and fever.

INTERDISCIPLINARY CARE



Ensuring adequate tissue oxygenation is the priority of care in treating anemia. Specific therapy is determined by the underlying cause of the disorder. Usual treatments include medications, dietary modifications, blood replacement, or supportive interventions. Table 34–1 outlines interdisciplinary care measures for selected types of anemia.

Diagnosis

When anemia is suspected, the following laboratory and diagnostic tests may be ordered:

- *Complete blood count (CBC)* is done to determine blood cell counts, hemoglobin, hematocrit, and red blood cell indices. The severity of the anemia, shape, volume, and iron content of the RBCs can help determine the cause of anemia.
- *Iron levels* and *total iron-binding capacity* are performed to detect iron deficiency anemia. A low serum iron concentration and elevated total iron-binding capacity are indicative of iron deficiency anemia.
- *Serum ferritin* is low due to depletion of the total iron reserves available for hemoglobin synthesis. Ferritin is an iron-storage protein produced by the liver, spleen, and bone marrow. Ferritin mobilizes stored iron when metabolic needs are higher than dietary intake.
- *Sickle cell test* is a screening test to evaluate hemolytic anemia and detect HbS.
- *Hemoglobin electrophoresis* separates normal hemoglobin from abnormal forms. It is used to evaluate hemolytic ane-

TABLE 34–1 Interdisciplinary Care Focus for Major Anemias

TYPE OF ANEMIA	INTERDISCIPLINARY CARE
Iron deficiency anemia	■ Increased dietary intake of iron-rich foods ■ Oral or parenteral iron supplements
Vitamin B_{12} deficiency	■ Increased dietary intake of foods containing vitamin B_{12} (e.g., meats, eggs, and dairy products) ■ Oral or parenteral vitamin B_{12} supplements ■ Parenteral vitamin B_{12} for deficiency due to malabsorption or lack of intrinsic factor
Folic acid deficiency	■ Increased dietary intake of foods rich in folic acid (folate) ■ Oral folic acid supplements ■ Folic acid supplements recommended for women who are pregnant or may become pregnant to prevent neural tube defects

Sickle cell anemia	■ Treatment is primarily supportive
	■ Hydroxyurea 10–30 mg/kg per day
	■ Sickle cell crisis:
	  ■ Rest
	  ■ Oxygen therapy to maintain SaO_2
	  ■ Narcotic analgesia
	  ■ Vigorous hydration
	  ■ Treatment of precipitating factors
	■ Acute chest syndrome:
	  ■ Careful hydration; hemodynamic monitoring
	  ■ Oxygen therapy
	  ■ Transfusion
	■ Folic acid supplements
	■ Blood transfusions during surgery or pregnancy as necessary
	■ Genetic counseling recommended

Thalassemia	■ Regular blood transfusions ■ Folic acid supplements ■ Possible splenectomy ■ Genetic counseling
Aplastic anemia	■ Withdrawal of the causative agent, if known ■ Blood transfusions ■ Bone marrow transplant as indicated

mia, diagnose thalassemia, and differentiate sickle cell trait from sickle cell disease.

- *Schilling test* measures vitamin B_{12} absorption before and after intrinsic factor administration to differentiate between pernicious anemia and intestinal malabsorption of the vitamin. A 24-hour urine sample is collected following administration of radioactive vitamin B_{12}. Lower than normal levels of the tagged B_{12} when intrinsic factor is given concurrently indicate malabsorption rather than pernicious anemia.
- *Bone marrow examination* is done to diagnose aplastic anemia. In aplastic anemia, normal marrow elements are significantly decreased as they are replaced by fat cells. Nursing implications for bone marrow collection are described in the Diagnostic Tests box in Chapter 33 ∞.
- *Quantitative assay of G6PD* may be performed to confirm a diagnosis of glucose-6-phosphate dehydrogenase deficiency.

Medications

Medications used to treat anemia depend on its cause. Iron replacement therapy is ordered for iron deficiency anemia. Supplemental iron may be given by mouth or parenterally. Intravenous administration of iron is becoming more common, particularly in clients with an acute deficiency and in anemia associated with chronic GI blood loss, chronic renal failure, and other chronic conditions that increase the need for blood cell production (e.g., cancers). The risk of anaphylaxis is a major concern when iron dextran is given intravenously. Other parenteral iron solutions, including intravenous sodium ferric gluconate (Ferrlecit) and iron sucrose (Venofer), carry a much lower risk of adverse and allergic reactions (Kasper et al., 2005).

Parenteral vitamin B_{12} is given when malabsorption or lack of intrinsic factor leads to vitamin B_{12} deficiency anemia. Folic acid is ordered for women of childbearing age, pregnant women, and clients with folic acid deficiency or sickle cell anemia to meet the increased demands of the bone marrow. Hydroxyurea, a drug that promotes fetal hemoglobin production, may be prescribed for clients with sickle cell disease, particularly those with frequent crises or severe disease. Resulting increased levels of fetal hemoglobin interfere with the sickling process and reduce the incidence of painful crises (Kasper et al., 2005). Nursing implications for clients receiving iron, vitamin B_{12}, and folic acid are found in the Medication Administration box on page 1112.

Erythropoietin may be ordered for clients with low erythropoietin levels (e.g., clients with chronic renal failure) and people with anemia associated with other chronic diseases. Erythropoietin is given subcutaneously, and may be given as often as three times a week in chronic renal failure. Because erythropoietin stimulates RBC production, adequate iron must be present. Clients receiving erythropoietin may require regular intravenous iron therapy as well (Kasper et al., 2005).

Immunosuppressive therapy with antithymocyte globulin (ATG), corticosteroids, and cyclosporine may be used to treat aplastic anemia. Androgens may stimulate blood cell production in some clients with aplastic anemia. See Chapter 13 ∞ for more information about immunosuppression.

Nutrition

Dietary modifications are recommended for nutritional deficiency anemias, such as iron deficiency anemia, vitamin B_{12} deficiency anemia, or folic acid deficiency anemia. Box 34–5 identifies good sources of dietary iron, vitamin B_{12}, and folic acid.

Blood Transfusion

Blood transfusions may be indicated to treat anemias resulting from major blood loss, such as from trauma or major surgery, and severe anemia regardless of cause. In acute hemorrhage, whole blood may be given to replace both blood cells and volume. A unit of packed red blood cells may be given when anemia is severe and the client demonstrates cardiovascular instability or compromise. Blood transfusions are fully discussed in Chapter 11 ∞.

Complementary Therapies

Complementary healthcare practitioners may recommend specific plant enzymes to treat nutritional anemias. Plant enzymes are believed to aid digestion of proteins, fats, and carbohydrates, facilitating absorption of their nutrients. Therapy is determined by the specific type of anemia. Plant enzymes should

BOX 34–5 Dietary Sources of Iron, Folic Acid, and Vitamin B_{12}

Iron

Iron in the diet comes from two sources. *Heme iron* makes up about one-half of the iron from animal sources. *Nonheme iron* includes the remaining iron from animal sources and all the iron from plants, legumes, and nuts. Heme iron promotes absorption of nonheme iron from other foods when both forms are consumed at the same time. Absorption of nonheme iron is also enhanced by vitamin C and inhibited by tea and coffee.

Sources of Heme Iron

- Beef
- Chicken
- Egg yolk
- Clams, oysters
- Pork loin
- Turkey
- Veal

Sources of Nonheme Iron

- Bran flakes
- Brown rice
- Whole-grain breads
- Dried beans
- Dried fruits
- Greens
- Oatmeal

Sources of Folic Acid

- Green leafy vegetables
- Broccoli
- Organ meats
- Eggs
- Wheat germ
- Asparagus
- Liver
- Milk
- Yeast
- Kidney beans

Sources of Vitamin B_{12}

- Liver
- Fresh shrimp and oysters
- Eggs
- Milk
- Kidney
- Meats (muscle)
- Cheese

MEDICATION ADMINISTRATION Drugs to Treat Anemia

IRON SOURCES

Ferrous sulfate (Feosol, Fer-in-sol)
Ferrous gluconate (Fergon, Ferralet, Fertinic)
Iron dextran injection (Imferon)
Iron polysaccharide
Iron sucrose (Venofer)
Sodium ferric gluconate (Ferrlecit)

Iron preparations are normally taken by mouth and are absorbed from the gastrointestinal tract. They are given to treat anemias resulting from iron deficiency or blood loss. When absorbed, iron combines with transferrin. This complex then is transported to the bone marrow and incorporated into hemoglobin.

Nursing Responsibilities

- Prior to giving the drug, assess for use of drugs that might interact with iron (e.g., antacids, allopurinol, chloramphenicol, tetracyclines, vitamin E), gastrointestinal bleeding, and manifestations of anemia.
- Administer iron preparations with orange juice to enhance absorption.
- If using an elixir, give it through a straw to prevent staining the teeth.
- Monitor for manifestations of iron toxicity: nausea, diarrhea, or constipation; symptoms of anaphylactic shock (extreme cases).
- Monitor hemoglobin and reticulocyte counts.
- If the client is also taking tetracyclines, schedule the dose of iron 2 hours before tetracycline (iron reduces the absorption of tetracycline).
- When administering IM or IV, monitor closely for anaphylaxis.

Health Education for the Client and Family

- Gastrointestinal side effects may be reduced by taking iron with food (but not milk, which decreases absorption).
- Stools may be dark green or black; this is harmless.
- Increase fluids and fiber in diet to decrease constipation.

VITAMIN B_{12} SOURCES

Cyanocobalamin (Kaybovite [oral], Anacobin [parenteral], Bedoz)

Cyanocobalamin is used to treat vitamin B_{12} deficiencies or malabsorption and pernicious anemia. It is rapidly absorbed when administered orally or by injection, and it is stored in the liver. Intrinsic factor is necessary for absorption from the gastrointestinal tract.

Nursing Responsibilities

- Do not expose crystalline injection to light.
- Assess for other drugs that might interfere with the therapeutic response: chloramphenicol, cimetidine, colchicine, and timed-release potassium decrease its effectiveness.
- Do not mix cyanocobalamin in a syringe with other medications.
- Administer parenteral doses intramuscularly or deep subcutaneously to decrease local irritation.
- Monitor hemoglobin, RBC counts, reticulocyte counts, and potassium levels.

Health Education for the Client and Family

- A burning sensation with injection is temporary.
- Avoid alcohol, which interferes with absorption.
- If used to treat pernicious anemia, the medication must be taken for life.

FOLIC ACID SOURCES

Folic acid (Folvite)

Synthetic folic acid is used to treat folic acid deficiency and megaloblastic or macrocytic anemia. It is absorbed from the gastrointestinal tract and stored in the liver.

Nursing Responsibilities

- Prior to giving the medication, assess for use of drugs that alter its effect: corticosteroids, methotrexate, oral contraceptives, phenytoin, sulfonamides.
- Do not mix folic acid with other medications in the same syringe.
- Monitor for possible hypersensitivity response of skin rash.

Health Education for the Client and Family

- Large doses of folic acid may cause the urine to become darker yellow.
- Excess alcohol intake increases folic acid requirements.

not be used alone to treat anemia, and it is important to check for possible interactions with prescribed medications before starting therapy.

NURSING CARE

For nursing care specific to the client with a nutritional anemia see the accompanying Nursing Care Plan.

Health Promotion

Nursing measures to prevent anemia focus on teaching good dietary habits to all clients, regardless of age. Stress the importance of consuming adequate amounts of iron, folate, and the B vitamins. Provide a list of dietary sources of these nutrients. Discuss alternate iron sources with vegetarian clients, and teach them that foods high in vitamin C enhance the absorption of iron from grains, legumes, and other sources. Emphasize the importance of adequate iron intake in women of childbearing age and older adults. Stress the increased need for these nutrients during pregnancy, and discuss strategies to ensure an adequate intake.

Assessment

Assessment data to collect for clients with suspected anemia includes:

- *Health history:* Complaints of shortness of breath with activity, fatigue, weakness, dizziness or fainting, palpitations; history of previous anemia, bleeding episodes; menstrual history (if appropriate); medications; chronic diseases; usual diet and patterns of alcohol intake or cigarette smoking.
- *Physical Examination:* General appearance, skin color; vital signs including temperature; heart and lung sounds; peripheral pulses, capillary refill; abdominal tenderness; obvious bleeding or bruising.

NURSING CARE PLAN A Client with Folic Acid Deficiency Anemia

Sheri Matthews is a 76-year-old widow who lives alone. She tells Lisa Apana, RN, the nurse in her care provider's office, that she liked to cook when her husband was alive, but preparing an entire meal just for herself seems senseless. She relates that her typical day's menu includes coffee for breakfast, a bologna sandwich and coffee for lunch, and a hot dog or two, a few cookies, and a glass of milk for dinner.

ASSESSMENT

Mrs. Matthews's nursing history includes a 20-lb (9-kg) weight loss since her husband died 8 months ago. She states that she sometimes has heart palpitations and always feels weak. Physical assessment shows: T 98.8°F (37.1°C), P 110, R 22, BP 90/52. Skin warm, pale, and dry. Diagnostic tests indicate folic acid deficiency anemia, and Mrs. Matthews is started on an oral folic acid supplement and instructed about foods containing folic acid.

DIAGNOSES

- *Activity Intolerance* related to weakness secondary to decreased tissue oxygenation
- *Imbalanced Nutrition: Less than Body Requirements* related to lack of motivation to cook and understanding of nutritional needs, as manifested by weight loss of 20 lb and folic acid deficiency
- *Deficient Knowledge* related to lack of information about a well-balanced diet and foods containing folic acid

EXPECTED OUTCOMES

- Verbalize the importance of taking folic acid supplements and eating a balanced diet.
- Gain at least 1 lb (0.45 kg) per week.
- Return to previous level of physical energy.
- Consume a balanced diet, including foods containing folic acid.

PLANNING AND IMPLEMENTATION

- Discuss foods required for a well-balanced diet, as well as dietary sources of folic acid.
- Develop a dietary plan with Mrs. Matthews that includes food preferences and foods that are easy and quick to prepare.
- Discuss the importance of taking the folic acid supplement. Advise to continue taking it even after she begins to feel better.
- Help Mrs. Matthews develop a schedule of activities that provides adequate rest and energy for cooking.

EVALUATION

Mrs. Matthews gained 1 lb (0.45 kg) during the first week of treatment. She has met with a nutritionist and has a better understanding of nutritional needs. She states that she can prepare hot meals when she schedules a rest period before and after lunch. Ms. Apana has provided written and verbal information about the folic acid supplement and diet. Mrs. Matthews verbalizes understanding, stating, "I will continue to take the folic acid until the doctor tells me to stop. I'm beginning to enjoy cooking again, now that I have a reason to cook!" Ms. Apana contacts the local senior services representative to determine if Mrs. Matthews is able to participate in the local Meals-on-Wheels program.

CRITICAL THINKING IN THE NURSING PROCESS

1. What is the pathophysiologic basis for Mrs. Matthews's abnormal vital signs during her initial assessment?
2. Design a week's menu that includes foods high in folic acid.
3. Why was Mrs. Matthews placed on a folic acid supplement in addition to dietary modifications?
4. Why is the older adult at increased risk for developing folic acid deficiency anemia? Consider physiologic, economic, and social factors.

See Evaluating Your Response in Appendix C.

- *Diagnostic tests:* CBC, hemoglobin, and hematocrit; bone marrow studies; specialized tests (e.g., hemoglobin electrophoresis, Schilling test).

Nursing Diagnoses and Interventions

Anemia affects circulating oxygen levels and tissue oxygenation. Priority nursing diagnoses include activity intolerance, altered oral mucous membranes, and self-care deficits. With acute blood-loss anemia, risk for insufficient cardiac output also is a priority. Clients with sickle cell disease have specific needs related to the effects of the disease on tissue perfusion; see the section on disseminated intravascular coagulation later in this chapter for nursing interventions appropriate to ineffective tissue perfusion, associated pain, and maintaining oxygenation.

Activity Intolerance

Anemia causes weakness and shortness of breath on exertion. These symptoms are due to decreased circulating oxygen levels secondary to low hemoglobin levels. Weakness, fatigue, and/or vertigo may occur even during activities of daily living, including those associated with self-care, home life, job performance, and social roles.

- Help identify ways to conserve energy when performing necessary or desired activities. *Modifying the approach to a particular activity may reduce cardiorespiratory symptoms and activity-related fatigue. Alternative ways of performing tasks (e.g., sitting when performing hygiene care and kitchen tasks) may reduce oxygen demands. In some cases, assistance from others is necessary to conserve energy and reduce symptoms.*
- Help the client and family establish priorities for tasks and activities. *Because family members may need to assume responsibility for additional tasks, the plan's success depends on mutually established goals.*
- Assist to develop a schedule of alternating activity and rest periods throughout the day. *Rest periods decrease oxygen needs, reducing strain on the heart and lungs, and allowing restoration of homeostasis before further activities.*
- Encourage 8 to 10 hours of sleep at night. *Rest decreases oxygen demands and increases available energy for morning activities.*
- Monitor vital signs before and after activity. *Vital signs provide a measure of activity tolerance. Increased heart and respiratory rates or a change in blood pressure may indicate intolerance of the activity.*

- Discontinue activity if any of the following occurs:
  a. Complaints of chest pain, breathlessness, or vertigo
  b. Palpitations or tachycardia that does not return to normal within 4 minutes of resting
  c. Bradycardia
  d. Tachypnea or dyspnea
  e. Decreased systolic blood pressure.

  These changes may signify cardiac decompensation due to insufficient oxygenation. The intensity, duration, or frequency of the activity needs to be reduced.
- Instruct the client not to smoke. *Smoking causes vasoconstriction and increases carbon monoxide levels in the blood, interfering with tissue oxygenation.*

Impaired Oral Mucous Membrane

Glossitis and cheilosis may occur with nutritional deficiencies of iron, folate, and vitamin B_{12}. The tongue and lips become very red, and fissures or cracks may form at the corners of the mouth.

- Monitor condition of lips and tongue daily. *Glossitis and cheilosis increase the risk for bleeding and infection and may require medical treatment. Pain and discomfort may interfere with oral intake, further worsening the nutritional deficiency.*
- Use a mouthwash of saline, saltwater, or half-strength peroxide and water to rinse the mouth every 2 to 4 hours. Avoid alcohol-based mouthwashes. *This cleanses and soothes oral mucous membranes. Alcohol-based mouthwashes further irritate and dry oral tissues.*
- Provide frequent oral hygiene (after each meal and at bedtime) with a soft bristle toothbrush or sponge. *Removing food debris from painful fissures promotes comfort. A soft toothbrush reduces irritation or bleeding of oral mucosa. Keeping the oral cavity clean also reduces the risk of infection.*
- Apply a petroleum-based lubricating jelly or ointment to the lips after oral care. *Lubricating ointment helps to retain moisture, facilitate healing, and protect the lips from other drying agents.*
- Instruct to avoid hot, spicy, or acidic foods. *Such foods may further irritate and dry mucous membranes.*
- Encourage soft, cool, bland foods. *Foods that are soothing to the mucous membranes promote comfort and help maintain adequate food and fluid intake. Minimizing oral pain may also promote compliance with oral care routines.*
- Encourage eating four to six small meals daily with high protein and vitamin content. *Small, frequent meals may be better tolerated, increasing intake. Nutrient-rich meals promote healing of the mucous membranes.*

Risk for Decreased Cardiac Output

Cardiac output may be affected by acute bleeding and volume loss or by heart failure resulting from severe anemia. In addition, impaired tissue oxygenation leads to an increased respiratory rate and dyspnea.

- Monitor vital signs, breath sounds, and apical pulse. *Increased cardiac workload can affect the blood pressure, heart, and respiratory rates. Increased blood flow can lead to heart murmur or abnormal heart sounds such as S_3 or S_4. Tachypnea and dyspnea may affect the depth of respirations, alveolar ventilation, and blood and tissue oxygenation.*
- Assess for pallor, cyanosis, and dependent edema. *Blood is shunted to the vital organs, causing vasoconstriction of skin vessels. This, in addition to lower levels of hemoglobin, causes pallor. Cyanosis, especially of the lips and nail beds, indicates inadequate oxygenation of blood. Dependent edema occurs in response to right ventricular failure.*

PRACTICE ALERT
Report signs of decreased cardiac output to the physician. Severe anemia can lead to heart failure, necessitating additional treatment.

- Closely monitor for manifestations of anaphylaxis (urticaria, erythema or flushing, edema, wheezing, dyspnea, nausea and vomiting, anxiety) when administering parenteral iron preparations, particularly iron dextran. Immediately notify the physician, and prepare to administer prescribed drugs such as diphenhydramine (Benadryl) or epinephrine as ordered. Institute cardiopulmonary resuscitation measures as necessary. *Anaphylaxis, a systemic type I hypersensitivity (allergic) reaction, is a risk when administering parenteral iron preparations, iron dextran in particular. Anaphylaxis can lead to severe cardiopulmonary compromise, necessitating emergency measures to preserve life.*

Self-Care Deficit

Energy expenditures for activities of daily living (ADLs) may cause oxygen demands to exceed supply in the client with severe anemia.

- Assist with ADLs, such as bathing, grooming, and eating, as needed. *Assistance decreases energy expenditures and tissue requirements for oxygen, reducing cardiac workload.*
- Discuss the importance of rest periods prior to such activities as dressing. *Rest reduces oxygen demand and cardiac workload. The person who is able to perform self-care in activities of daily living maintains independence, self-esteem, and morale.*

NANDA, NIC, and NOC Linkages

See Chart 34–1 for linkages between NANDA nursing diagnoses, nursing interventions, and nursing outcomes for the client with anemia.

Community-Based Care

With the exception of anemia resulting from acute hemorrhage, most clients with anemia are treated in the home and community setting. Include the following topics when preparing the client and family for home care:

- Nutritional strategies to address deficiencies
- Prescribed medications, vitamins, or mineral supplements and their appropriate use, intended effect, possible adverse effects, and interactions with food or other medications
- Energy conservation strategies
- Other recommended treatment measures and follow-up
- If the anemia is genetically transmitted, such as sickle cell anemia, include inheritance patterns of the disorder, symptoms of crisis, and manifestations to report to the physician.

Provide referrals for counseling to facilitate decisions about pregnancy as indicated. Also refer for nutritional assistance and

NANDA, NIC, AND NOC LINKAGES
CHART 34–1 **The Client with Anemia**

NANDA

Activity Intolerance

↓

NIC

Energy Management

Environmental Management

Nutrition Management

↓

NOC

Activity Tolerance

Endurance

Energy Conservation

Data from *NANDA's Nursing Diagnoses: Definitions & Classification 2005–2006* by NANDA International (2005), Philadelphia; *Nursing Interventions Classification (NIC)* (4th ed.) by J. M. Dochterman & G. M. Bulechek (2004), St. Louis, MO: Mosby; and *Nursing Outcomes Classification (NOC)* (3rd ed.) by S. Moorhead, M. Johnson, and M. Mass (2004), St. Louis, MO: Mosby.

teaching, home health care, or assistance with self-care and home maintenance activities as indicated. Older adults with nutritional anemias may benefit from community services such as senior meals or Meals-on-Wheels.

THE CLIENT WITH MYELODYSPLASTIC SYNDROME

Myelodysplastic syndrome (MDS) is a group of blood disorders characterized by abnormal-appearing bone marrow and cytopenia (low numbers of circulating blood cells). MDS is not a single disease; at least five variations of the disorder have been identified. Anemia that does not respond to treatment (*refractory anemia*) is a characteristic of most forms of myelodysplasia.

Idiopathic MDS primarily affects older adults; men have a slightly higher incidence of the disorder than women. Risk factors for secondary MDS include exposure to environmental toxins such as cigarette smoke, benzene, radiation, radiation therapy or chemotherapy for cancer treatment, and other anemias such as aplastic anemia or Fanconi's anemia (Demakos & Linebaugh, 2005; Kasper et al., 2005).

FAST FACTS

- Idiopathic or primary MDS accounts for 70% to 80% of all identified cases.
- Twenty percent to 30% of MDS cases occur as a secondary condition, related to factors such as smoking or exposure to environmental toxins, radiation, chemotherapy, or other risk factors (Demakos & Linebaugh, 2005).

Pathophysiology

MDS is a stem cell disorder in which stem cells fail to reproduce and differentiate into the various types of blood cells. The genetic components of stem cells (nuclear DNA and/or mitochondrial DNA) are altered. The bone marrow loses its ability to produce normal blood cells, instead producing abnormal (*dysplastic*) cells. With significant alterations, leukemia (proliferation of abnormal white blood cells) may develop in people with MDS.

Manifestations

Anemia is the predominant early manifestation of MDS. The client may develop symptoms of the anemia with increasing fatigue, weakness, dyspnea and pallor. In many cases, the disorder is asymptomatic, identified when a routine blood count shows anemia. Splenomegaly may develop, leading to discomfort and a feeling of fullness in the left upper quadrant of the abdomen. Hepatomegaly also may develop, leading to right upper quadrant discomfort. Thrombocytopenia can lead to abnormal bleeding tendencies, and neutropenia increases the risk for infection (Demakos & Linebaugh, 2005).

INTERDISCIPLINARY CARE

Clients with MDS require long-term supportive care and therapy to maintain their quality of life. Stem cell transplant offers the only real hope for cure in MDS. See the Interdisciplinary Care section of the client with leukemia later in this chapter for more information about stem cell transplant and associated nursing care.

Diagnosis

- The *CBC* reveals anemia. Although anemia may be the only abnormality of the blood count, the white blood cell (WBC) count also may be low, as may be the platelet count. Abnormalities of size and shape may be noted in all blood cells.
- The *bone marrow* often appears normal, although precursor cells may have an abnormal appearance. Increased numbers of myeloblasts (granulocyte precursor cells) may be present in the bone marrow.
- *Serum erythropoietin, vitamin B_{12}, serum iron, total iron-binding capacity, ferritin levels,* and *RBC folate levels* are drawn to help guide supportive therapy.

Treatment

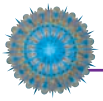
Management of MDS is based on the severity of the disease. Several classification systems are available, including the French-American-British (FAB) classification system, the International Prognostic Scoring System (IPSS), and the World Health Organization (WHO) classification system (National Comprehensive Cancer Network [NCCN], 2006). These systems are used to guide therapy for the client with MDS.

All clients with MDS require monitoring, with regular physician visits and laboratory evaluations. Psychosocial support is provided to assist the client and family dealing with a chronic, progressive, and ultimately fatal disease.

Clients with MDS may require frequent red blood cell transfusions to treat the predominant anemia. Each unit of packed

RBCs contains 250 to 300 mg of iron. The body is unable to excrete this excess iron, so it accumulates, leading to problems such as endocrine dysfunction, cirrhosis, pericarditis, and heart failure. *Iron chelation therapy* is used to remove excess iron from the body. Desferrioxamine (Desferal) is administered by slow intravenous infusion or continuous subcutaneous infusion using an infusion pump to maintain a normal or negative iron balance. This drug is relatively safe, although local skin reactions such as rash and urticaria may develop. An oral form of the drug, deferasirox, is available, but not widely used.

Blood cell growth factors may be administered to stimulate stem cell development in MDS, although the response rate is low. Platelet transfusions are given when bleeding occurs due to low platelet levels. Antibiotic therapy is initiated for bacterial infections (NCCS, 2006). Chemotherapy regimens similar to those employed to treat leukemia may be used, but rarely are effective in treating MDS. Azacitidine (Vidaza), an antileukemic agent that acts on abnormal blood-forming cells in the bone marrow, may be more effective in treating MDS than standard chemotherapy regimens (Demakos & Linebaugh, 2005). As previously noted, stem cell transplant offers the only hope for cure. This high-risk therapy, however, is reserved for higher risk clients. Factors such as age, functional ability, and other existing disease conditions help guide the decision to undergo stem cell transplant (NCCN, 2006).



NURSING CARE

Nursing Diagnoses and Interventions

Activity intolerance and the need for education about this disorder are the priorities of nursing care for the client with MDS being managed in a community-based setting. Although neutropenia and thrombocytopenia may accompany the anemia of MDS, these problems are less common. See the section of this chapter on leukemia for additional potential nursing diagnoses and interventions for the client with MDS.

Activity Intolerance

The client with MDS experiences fatigue, weakness, and shortness of breath on exertion related to the lack of RBCs and ineffective oxygen transport. These symptoms may affect the client's ability to maintain self-care, home life, job performance, and social roles.

- Monitor vital signs, breath sounds, and apical pulse. *Increased cardiac workload due to anemia and impaired oxygen transport can affect the blood pressure, heart, and respiratory rates. Increased blood flow can lead to heart murmur or abnormal heart sounds such as S_3 or S_4. Accumulated iron can lead to pericarditis and a pericardial friction rub.*
- Help identify energy-conserving ways of performing necessary or desired activities. *Alternative ways of performing tasks (e.g., sitting while performing hygiene measures) may reduce oxygen demands and fatigue.*
- Help the client and family establish priorities for tasks and activities. *Because family members may need to assume responsibility for additional tasks, the plan's success depends on mutually established goals.*
- Suggest planning recreational activities following a transfusion and adjusting activity level between transfusions to match energy and minimize fatigue. *The client with MDS will have more energy and activity tolerance following a transfusion when RBC counts, hemoglobin, and hematocrit approach normal levels and oxygen transport is optimal.*
- Encourage 8 to 10 hours of sleep at night. *Rest decreases oxygen demands and increases available energy for morning activities.*
- Discontinue activity if any of the following occurs:
  a. Complaints of chest pain, breathlessness, or vertigo
  b. Palpitations or tachycardia that does not return to normal within 4 minutes of resting
  c. Bradycardia
  d. Tachypnea or dyspnea
  e. Decreased systolic blood pressure.

  These changes may signify cardiac decompensation due to insufficient oxygenation. The intensity, duration, or frequency of the activity needs to be reduced.
- Instruct the client not to smoke. *Smoking causes vasoconstriction and increases carbon monoxide levels in the blood, interfering with tissue oxygenation.*

Risk for Ineffective Health Maintenance

MDS is a chronic, usually progressive disorder, requiring active management to maintain functional status and quality of life. Regular visits to the physician or clinic may be necessary. In addition, the client or family members may need to learn to administer iron chelation therapy or chemotherapy drugs and measures to prevent complications. The chronic nature of the disorder and the often advanced age of the client and family caregivers may interfere with effective management of the disorder.

- Assess knowledge of the disorder and the related treatments. *Assessment allows identification of knowledge gaps and provides a basis on which to provide additional information. Impaired disease management may be due to lack of knowledge or an inability to learn and perform psychomotor skills (e.g., administration of parenteral drug therapy).*
- Provide information about the disorder, its effects, and prescribed medications and treatments. *Individualized instruction is more effective than general, possibly irrelevant information. The client and caregivers need to be able to identify and manage possible adverse effects of drug therapy, as well as recognize potential complications to be reported to the physician.*
- Provide emotional support, expressing confidence in the client's and caregivers' abilities to manage care. *Emotional support helps the client and family caregivers incorporate the care regimen into their lifestyle.*
- Provide supervised learning and practice opportunities for administering parenteral medications if ordered. *Successful practice sessions instill confidence in the ability to manage care and provide an opportunity for questions and exploring alternatives.*

Community-Based Care

The client with myelodysplastic syndrome needs information about this chronic and ultimately fatal disease. Provide information about treatment options, including management of the infusion pump if ordered. Discuss the timing of and options for stem cell transplant, and assist the client to evaluate the potential benefits and risks of this treatment option.

THE CLIENT WITH POLYCYTHEMIA

Polycythemia, or *erythrocytosis,* is an excess of red blood cells characterized by a hematocrit higher than 55%. The two major types of polycythemia are primary and secondary. A third type of polycythemia, relative polycythemia, results from a fluid volume deficit, not excess RBCs.

FAST FACTS

- *Primary polycythemia (polycythemia vera*) is uncommon.
  - In primary polycythemia, RBC production is increased.
  - Primary polycythemia more commonly affects men of European Jewish ancestry between age 40 and 70.
- *Secondary polycythemia* (*erythrocytosis*) is the most common form of polycythemia.
  - Secondary polycythemia occurs when erythropoietin levels are elevated.
  - It may affect clients of any age or origin.
  - It usually develops in response to hypoxia (living at a high altitude, smoking, or chronic lung disease).
- *Relative polycythemia* occurs due to fluid deficit, not excess RBCs.
  - In relative polycythemia the total RBC count is normal.
  - The hematocrit is elevated because of increased cell concentration.
  - It is corrected by rehydration.

Pathophysiology

Primary Polycythemia

Primary polycythemia, or polycythemia vera (PV), is a neoplastic stem cell disorder characterized by overproduction of RBCs and, to a lesser extent, white blood cells and platelets. It is classified as a myeloproliferative disorder. Its cause is unknown. In PV, colonies of endogenous erythroid stem cells develop. These colonies produce RBCs in the absence of erythropoietin, leading to excess RBC production.

MANIFESTATIONS Initially, PV is asymptomatic, and the diagnosis may be made during routine blood tests. Its manifestations are caused by increased blood volume and viscosity. Hypertension is common, and may lead to complaints of headaches, dizziness, and vision and hearing disruptions. Venous stasis causes *plethora,* a ruddy, red color of the face, hands, feet, and mucous membranes. This often is accompanied by severe, painful itching of the fingers and toes. Retinal and cerebral vessels may be engorged. Hypermetabolism develops, causing weight loss and night sweats. Mental status may be altered, leading to drowsiness or delirium.

Thrombosis and hemorrhage are potential complications of PV. Thrombosis may cause transient ischemic attacks, angina, or manifestations of peripheral vascular disease. Gastrointestinal bleeding may occur, and portal hypertension may develop.

Secondary Polycythemia

Secondary polycythemia, or erythrocytosis, is increased numbers of RBCs in response to excess erythropoietin secretion or prolonged hypoxia. Secondary polycythemia is the most common form of polycythemia.

Abnormally high erythropoietin levels can result from kidney disease or erythropoietin-secreting tumors (e.g., renal cell carcinoma). Chronic hypoxia that stimulates erythropoietin release is a more common cause of secondary polycythemia. People living at high altitudes where the atmospheric oxygen pressure is lower develop a degree of polycythemia, as do people with chronic heart or lung disease and smokers. Abnormal hemoglobin that forms tighter bonds with oxygen also may lead to secondary polycythemia.

MANIFESTATIONS The manifestations of secondary polycythemia are similar to those of primary polycythemia. Splenomegaly, however, does not develop. Early symptoms often are overshadowed by the manifestations of the underlying disorder. For the manifestations of polycythemia see the box below.

INTERDISCIPLINARY CARE

Diagnosis

In PV, serum erythropoietin levels are low. Bone marrow studies show hyperplasia of all hematopoietic elements. With secondary polycythemia, serum erythropoietin levels usually are high, and bone marrow studies show only red stem cell hyperplasia.

Treatments

For secondary polycythemia, treatment focuses on the underlying cause of the disorder. It is a physiologic response in people living at high altitudes, and unless the hematocrit is too high or oxygen saturation levels are low, no treatment is usually necessary. Smokers are urged to quit. Measures to raise oxygen saturation levels and reduce tissue hypoxia often will relieve the polycythemia. Clients with both primary and secondary polycythemia benefit from periodic phlebotomy, removing 300 to 500 mL of blood, to keep blood volume and viscosity within normal levels. For PV, chemotherapeutic agents such as hydroxyurea may be used to suppress marrow function but may

MANIFESTATIONS of Polycythemia

- Hypertension
- Headache, tinnitus, blurred vision
- Plethora: dark redness of the lips, feet, ears, fingernails, and mucous membranes
- Splenomegaly (polycythemia vera)
- Severe pruritus, extremity pain
- Weight loss, night sweats
- Gastrointestinal bleeding
- Intermittent claudication
- Symptoms from thrombosis within various organs

increase the risk of developing leukemia (discussed later in this chapter). Pruritus may be relieved by antihistamines, or may require more aggressive treatment with interferon alpha or other treatments. One 325-mg aspirin tablet daily may be ordered to control thrombosis without increasing the risk of bleeding.

NURSING CARE

Preventing polycythemia begins with educating children and adults about the dangers of smoking. Measures to reduce risk factors for cardiovascular disease also may be beneficial.

This chronic condition is managed in community-based settings unless a complication develops. Teach the client and family the importance of maintaining adequate hydration, and increasing fluid intake during hot weather and when exercising. Discuss measures to prevent blood stasis: elevating legs and feet when sitting, using support stockings, and continuing treatment measures. Instruct to report manifestations of thrombosis (leg or calf pain, chest pain, neurologic symptoms) or bleeding (black, tarry stools, vomiting of blood or coffee-grounds emesis) immediately. Monitor the hematocrit and cell counts throughout treatment.

Examples of nursing diagnoses appropriate for the client with polycythemia follow:

- *Decisional Conflict Regarding Smoking Cessation* related to addictive effects
- *Pain* related to effects of altered blood flow in distal extremities
- *Risk for Ineffective Tissue Perfusion* related to sluggish blood flow and increased risk for thrombosis.

WHITE BLOOD CELL AND LYMPHOID TISSUE DISORDERS

Disorders of the white blood cells and lymphoid tissue include infectious mononucleosis, the leukemias, multiple myeloma, and malignant lymphomas (Hodgkin's disease and non-Hodgkin's lymphoma). Review the physiology of WBCs and lymphoid tissues and assessment of their function in Chapter 33 ∞ before proceeding with this section.

THE CLIENT WITH LEUKEMIA

Leukemia (literally, "white blood") is a group of chronic malignant disorders of white blood cells and white blood cell precursors. In leukemia, the usual ratio of red to white blood cells is reversed. Leukemias are characterized by replacement of bone marrow by malignant immature white blood cells, abnormal immature circulating WBCs, and infiltration of these cells into the liver, spleen, and lymph nodes throughout the body.

Incidence and Risk Factors

Although leukemia is often thought of as a childhood disease, it is diagnosed 10 times more often in adults than in children. An estimated 34,810 new cases of leukemia occur annually; slightly more than half are acute leukemia and less than half are chronic leukemia. In 2005, the ACS (2005) estimated that approximately 22,570 people died of leukemia. The highest incidence of leukemia is found in the United States, Canada, Sweden, and New Zealand (McCance & Huether, 2006).

Although the cause of most leukemias is unknown, certain risk factors have been identified. Men are affected more frequently than are women. People with certain genetic disorders such as Down syndrome have a higher incidence of leukemia. Environmental risk factors play a role as well. Risk factors for myeloid leukemia include cigarette smoking and chemicals such as benzene (present in cigarette smoke and gasoline). Exposure to ionizing radiation increases the risk for several types of leukemia. Clients who have undergone treatment for cancer have an increased risk. The human T-cell leukemia/lymphoma virus-1, a retrovirus, is known to cause certain leukemias and lymphomas (ACS, 2005).

Physiology Review

White blood cells are the most diverse of the cellular components of the blood. White blood cells arise from three different precursor cells: myeloblasts, which further differentiate into the granular leukocytes (granulocytes), neutrophils, eosinophils, and basophils; monoblasts, which mature into circulating monocytes, and ultimately into macrophages; and lymphoblasts, which become lymphocytes and mature in lymphoid tissue to B cells and T cells.

As a whole, the primary function of WBCs is to help maintain the body's immune defenses. Neutrophils, the most numerous WBC in circulation, are active phagocytes, the first cells to arrive to injured tissue. Monocytes and macrophages also are phagocytic cells that dispose of foreign and waste material from tissues. Eosinophils and basophils are more specialized. Eosinophils are primarily involved in allergic responses and parasitic infections. Basophils are actively involved in the inflammatory response, releasing substances such as histamine and heparin into inflamed tissues. Lymphocytes, the smallest of the WBCs, are an integral part of the immune system. B cells are part of the humoral immune response, producing antibodies to specific antigens. T cells are part of the cell-mediated immune response. For more information about the inflammatory and immune responses, see Chapter 12 ∞. The normal WBC count and differential are presented in Table 34–2.

TABLE 34–2 Normal White Blood Cell Count and Differential

LABORATORY TEST	VALUE
WBC count	5000–10,000/mm^3
Differential WBC count	
Neutrophils	60–70% or 3000–7000/mm^3
Eosinophils	1–3% or 50–400/mm^3
Basophils	0.3–0.5% or 25–200/mm^3
Lymphocytes	20–30% or 1000–4000/mm^3
Monocytes	3–8% or 100–600/mm^3

Pathophysiology

Leukemia begins with malignant transformation of a single stem cell. Leukemic cells proliferate slowly, but do not differentiate normally. They have a prolonged life span and accumulate in the bone marrow. As they accumulate, they compete with the proliferation of normal cells. Leukemic cells do not function as mature WBCs, and are ineffective in the inflammatory and immune processes. Leukemic cells replace normal hematopoietic elements in the marrow. Because erythrocyte- and platelet-producing cells are crowded out, severe anemia, splenomegaly, and bleeding difficulties result.

Leukemic cells leave the bone marrow and travel through the circulatory system, infiltrating other body tissues such as the CNS, testes, skin, gastrointestinal tract, and the lymph nodes, liver, and spleen. Death usually is due to internal hemorrhage and infections.

Manifestations

The general manifestations of leukemia (regardless of type) result from anemia, infection, and bleeding. These include pallor, fatigue, tachycardia, malaise, lethargy, and dyspnea on exertion. Infection may cause fever, night sweats, oral ulcerations, and frequent or recurrent respiratory, urinary, integumentary, or other infections. Increased bleeding due to thrombocytopenia leads to bruising, petechiae, bleeding gums, and bleeding within specific organs and tissues. *Multisystem Effects of Leukemia* can be seen on page 1120.

Other manifestations result from leukemic cell infiltration, increased metabolism, and increased leukocyte destruction. Infiltration of the liver, spleen, lymph nodes, and bone marrow causes pain and tissue swelling in the involved areas. Meningeal infiltration may cause manifestations of increased intracranial pressure, such as headache, altered level of consciousness, cranial nerve impairment, nausea, and vomiting. Infiltration of the kidneys may affect renal function, with decreased urine output and increased blood urea nitrogen and creatinine. Increased metabolism causes heat intolerance, weight loss, dyspnea on exertion, and tachycardia. Destruction of large numbers of WBCs releases substantial amounts of uric acid into the circulation; uric acid crystals may obstruct renal tubules, causing renal insufficiency.

Without treatment, leukemia is invariably fatal, usually due to complications of leukemic cell infiltration of bone marrow or vital organs. With treatment, prognosis varies. The overall 5-year survival rate is 46%. Survival rates differ by type of leukemia: People with acute myeloid leukemia have a 20% 5-year survival rate, whereas the rate is 73% for people with chronic lymphocytic leukemia (American Cancer Society, [ACS], 2005). The types, pathology, manifestations, and treatment for the major leukemias are outlined in Table 34–3.

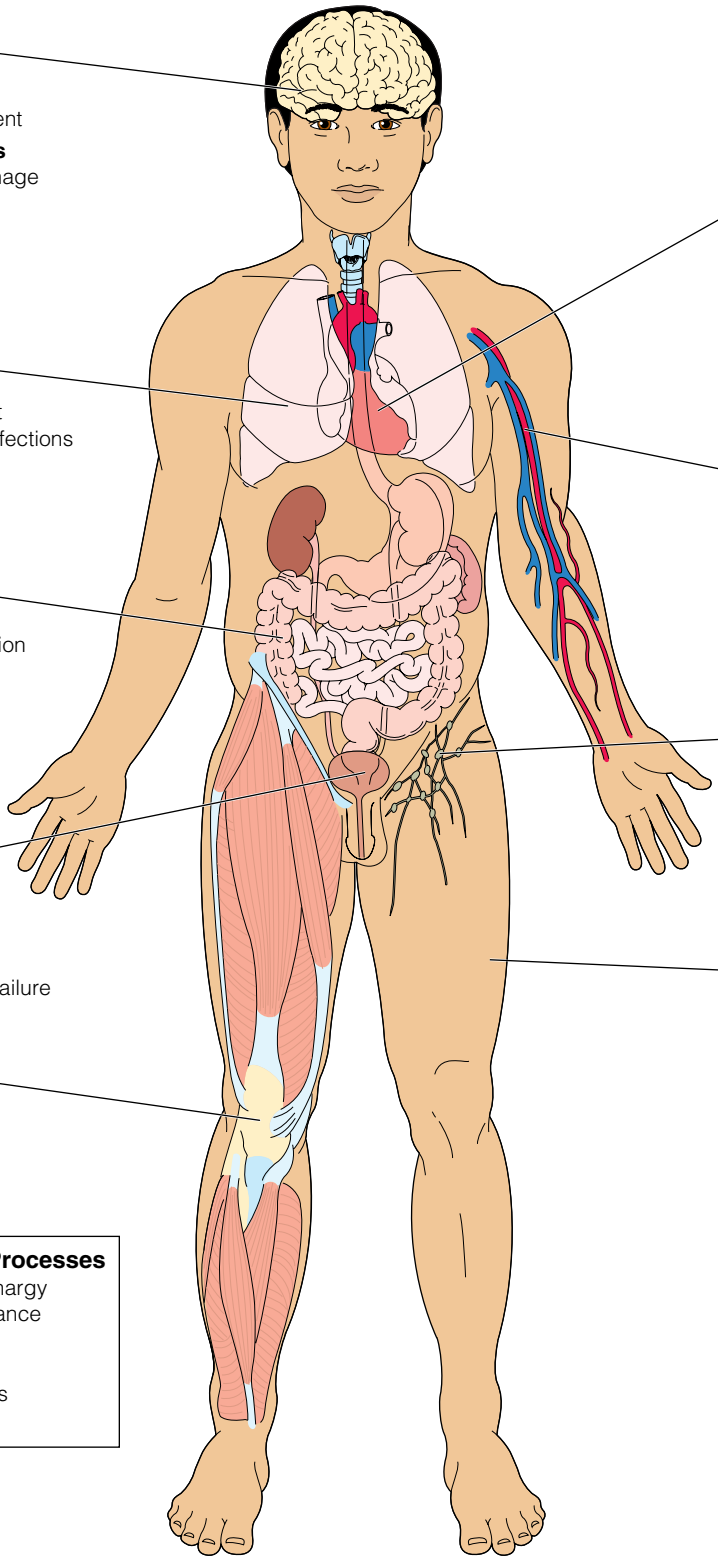
Classifications

Leukemias are classified by their acuity and by the predominant cell type involved. The *acute* leukemias are characterized by an acute onset, rapid disease progression, and immature or undifferentiated blast cells. *Chronic* leukemias, on the other hand, have a gradual onset, prolonged course, and abnormal mature-appearing cells. *Lymphocytic* (or *lymphoblastic*) leukemias involve immature lymphocytes and their precursor cells in the bone marrow. Lymphocytic leukemias infiltrate the spleen, lymph nodes, CNS, and other tissues. *Myeloid* (also called *myelogenous, myelocytic,* or *myeloblastic*) leukemias involve myeloid stem cells in the bone marrow, interfering with the maturation of all types of blood cells, including granulocytes, RBCs, and thrombocytes (Porth, 2005). Acute lymphoblastic leukemia is the most common type of leukemia in children. In adults, acute myeloid leukemia and chronic lymphocytic leukemia are the most common types (McCance &

TABLE 34–3 **Major Types of Leukemia**

CLASSIFICATION	CHARACTERISTICS	MANIFESTATIONS	TREATMENT
Acute lymphoblastic leukemia (ALL)	Primarily affects children and young adults; leukemic cells may infiltrate CNS	Recurrent infections; bleeding; pallor, bone pain, weight loss, sore throat, fatigue, night sweats, weakness	Chemotherapy; bone marrow transplant (BMT), or stem cell transplant (SCT)
Chronic lymphocytic leukemia (CLL)	Primarily affects older adults; insidious onset and slow, chronic course	Fatigue; exercise intolerance; lymphadenopathy and splenomegaly; recurrent infections, pallor, edema, thrombophlebitis	Often requires no treatment; chemotherapy; BMT
Acute myeloid leukemia (AML)	Common in older adults, may affect children and young adults. Strongly associated with toxins, genetic disorders, and treatment of other cancers	Fatigue, weakness, fever; anemia; headache; bone and joint pain; abnormal bleeding and bruising; recurrent infection; lymphadenopathy, splenomegaly, and hepatomegaly	Chemotherapy; SCT
Chronic myeloid leukemia (CML)	Primarily affects adults; early course slow and stable, progressing to aggressive phase in 3–4 years	*Early:* weakness, fatigue, dyspnea on exertion; possible splenomegaly *Later:* fever, weight loss, night sweats	Interferon alpha; chemotherapy with imatinib mesylate (Gleevec), SCT

MULTISYSTEM EFFECTS **of Leukemia**



Neurologic
- Headache
- Altered LOC
- Cranial nerve impairment

Potential complications
- Subarachnoid hemorrhage
- Retinal hemorrhage
- Seizures, coma

Respiratory
- Dyspnea on exertion
- Pharyngitis, sore throat
- Frequent respiratory infections

Potential complication
- Pulmonary bleeding

Gastrointestinal
- Anorexia, nausea
- Oral ulcerations, infection
- Bleeding gums
- Gingival hyperplasia (gum overgrowth)
- Abdominal pain
- Hepatomegaly
- Occult GI bleeding

Urinary
- Urinary tract infection
- Hematuria

Potential complication
- Renal insufficiency or failure

Musculoskeletal
- Weakness
- Bone tenderness, pain
- Joint pain

Cardiovascular
- Tachycardia, palpitations
- Orthostatic hypotension
- Heart murmurs
- Hematomas
- Edema

Potential complications
- Hemorrhage
- Thrombophlebitis

Hematologic
- Anemia
- Thrombocytopenia
- Leukopenia
- Bleeding (epistaxis)
- Splenomegaly

Potential complication
- DIC

Immunologic
- Frequent or recurrent infections
- Lymphadenopathy

Potential complications
- Abscesses
- Septicemia

Integumentary
- Skin and mucous membrane pallor
- Petechiae
- Bruising, purpura
- Ulcerations
- *Chloromas* (skin infiltrations near bony prominences)

Metabolic Processes
- Malaise, lethargy
- Heat intolerance
- Diaphoresis
- Chills, fever
- Night sweats
- Weight loss

Huether, 2006). In summary, the general types of leukemia are as follows:

- Acute lymphocytic (lymphoblastic) leukemia (ALL)
- Chronic lymphocytic leukemia (CLL)
- Acute myeloid (myeloblastic) leukemia (AML)
- Chronic myeloid (myelogenous) leukemia (CML).

This general system of classifying leukemias does not differentiate subtypes of acute leukemias. The FAB system for classifying acute leukemias further differentiates acute leukemias by the predominant cell involved and the degree of cell differentiation (Table 34–4).

Acute Myeloid Leukemia

Acute myeloid leukemia is characterized by uncontrolled proliferation of myeloblasts (the precursors of granulocytes) and hyperplasia of the bone marrow and spleen (Figure 34–5 ■). AML accounts for 80% of acute leukemia cases in adults (Copstead & Banasik, 2005). Treatment induces complete remission in 66% of clients, although only about 30% to 40% achieve cure or long-term remission (Porth, 2005).

The manifestations of AML result from neutropenia and thrombocytopenia. Decreased neutrophils lead to recurrent severe infections, such as pneumonia, septicemia, abscesses, and mucous membrane ulceration. The manifestations of thrombocytopenia include petechiae, purpura, ecchymoses (bruising), epistaxis (nosebleeds), hematomas, hematuria, and gastrointestinal bleeding. Bone infarctions or subperiosteal infiltrates of leukemic cells may cause bone pain. Anemia is a late manifestation, causing fatigue, headaches, pallor, and dyspnea on exertion. Death usually results from infection or hemorrhage.

Bone marrow aspiration shows a proliferation of immature WBCs. The CBC shows thrombocytopenia and normocytic, normochromic anemia.

Chronic Myeloid Leukemia

Chronic myeloid leukemia is characterized by abnormal proliferation of all bone marrow elements. This type of leukemia constitutes approximately 15% of adult leukemias. It affects men more frequently than women. The onset of CML typically is between ages 30 or 40 and 50, although it is seen in children and adolescents as well (Copstead & Banasik, 2005; Porth 2005).

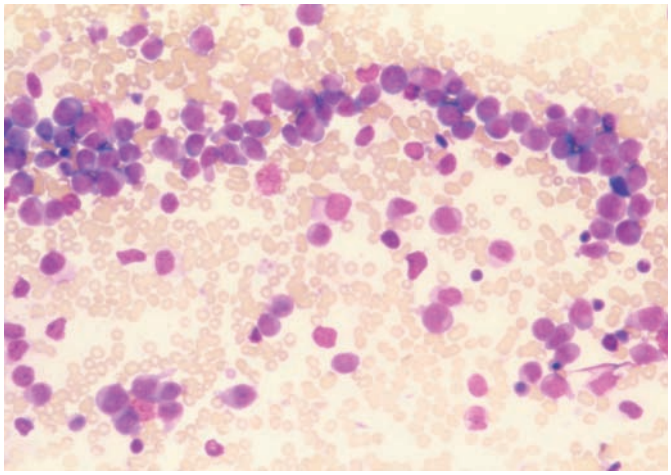


Figure 34–5 ■ A blood smear from the bone marrow of a client with acute myeloid leukemia. Note the abnormally large number of myelocyte WBCs (stained purple) among the small RBCs.

Source: Dr. Gopal Murti, Photo Researchers, Inc.

CML is usually associated with a chromosome abnormality called the Philadelphia chromosome, a balanced translocation of chromosome 22 to chromosome 9 (Figure 34–6 ■). The fusion gene produced by this translocation, known as *bcr/abl,* is an *oncogene* capable of initiating a malignancy. Very large doses of ionizing radiation also may induce CML in some clients (Kasper et al., 2005).

People with CML are often asymptomatic in the early stages and, in fact, are often diagnosed when a routine blood test reveals abnormal cell counts. Anemia causes weakness, fatigue, and dyspnea on exertion. The spleen often is enlarged, causing abdominal discomfort. Within 3 to 4 years, disease progresses to a more aggressive phase. Rapid cell proliferation and hypermetabolism cause fatigue, weight loss, sweating, and heat intolerance. The spleen enlarges, leading to a sensation of abdominal fullness and discomfort. Platelet function is affected in this stage, leading to bleeding and increased bruising. Finally, the disease evolves to acute leukemia, with blast cell proliferation. This stage, known as the *terminal blast crisis phase,* is characterized by significant constitutional manifestations, splenomegaly, and infiltration of leukemic cells into the skin, lymph nodes, bones, and CNS

TABLE 34–4 **FAB Classification of Acute Leukemia**

TYPE	CLASS	PREDOMINANT CELLS	PROGNOSIS
Acute lymphocytic leukemia	L_1	Immature lymphoblasts	>90% remission rate in children
	L_2	Mature lymphoblasts	Relapse common after 2 or more years of remission
Acute myeloid leukemia	M_0	Undifferentiated cells	Poor
	M_1	Immature myeloblasts	Good; complete response in 65% or more
	M_2	Mature myeloblasts	Good for 2 or more years of remission
	M_3	Promyelocytes	Good in adults
	M_4	Myelocytes and monocytes	Poorest in adults
	M_5	Poorly or well-differentiated monocytes	Poor
	M_6	Predominant erythroblasts	Variable
	M_7	Megakaryocytes	

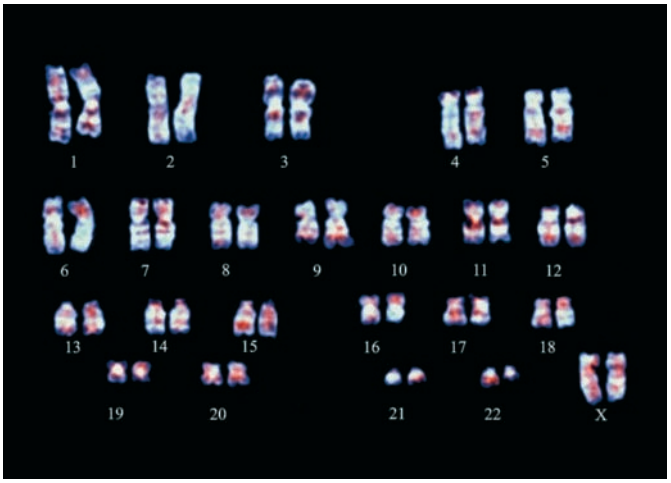


Figure 34–6 ■ The Philadelphia chromosome. Note the chromosomes of pairs 9 and 22. In each instance, the left-hand chromosome of the pair is normal, whereas an exchange of material between chromosomes has made the right-hand chromosome 9 larger and the right-hand chromosome 22 smaller. In stem cells within the bone marrow, the chromosome 22 defect leads to chronic myeloid leukemia.

Source: Addenbrookes Hospital, Photo Researchers, Inc.

(Porth, 2005). Survival following the onset of this final stage averages only 2 to 4 months.

Acute Lymphocytic Leukemia

Acute lymphocytic leukemia is the most common type of leukemia in children and young adults. In adults, ALL is rarely seen until late middle age, and then its incidence increases with aging. Genetic factors may play a role in its development, particularly the *bcr/abl* translocation also implicated in CML (Copstead & Banasik, 2005).

Most (80%) cases of ALL result from malignant transformation of B cells, with the remaining 20% arising from T cells. The malignant cells resemble immature lymphocytes (*lymphoblasts*); however, they do not mature or function effectively to maintain immunity. These lymphoblasts accumulate in the bone marrow, lymph nodes, and spleen, as well as in circulating blood. Some types of lymphoma (discussed later in this chapter) are thought to represent a later stage of the same disease.

The onset of ALL is usually rapid. Lymphoblasts proliferating in bone marrow and peripheral tissues crowd the growth of normal cells (Figure 34–7 ■). Normal hematopoiesis is suppressed, leading to thrombocytopenia, leukopenia, and anemia. Manifestations of infections, bleeding, and anemia develop. Bone pain resulting from rapid generation of marrow elements, lymphadenopathy, and liver enlargement are also common. Infiltration of the CNS causes headaches, visual disturbances, vomiting, and seizures.

The CBC shows an elevated WBC count with increased lymphocytes on the differential. RBC and platelet counts are decreased. Bone marrow studies reveal a hypercellular marrow with growth of lymphoblasts. Combination chemotherapy produces complete remission in 80% to 90% of adults with ALL.

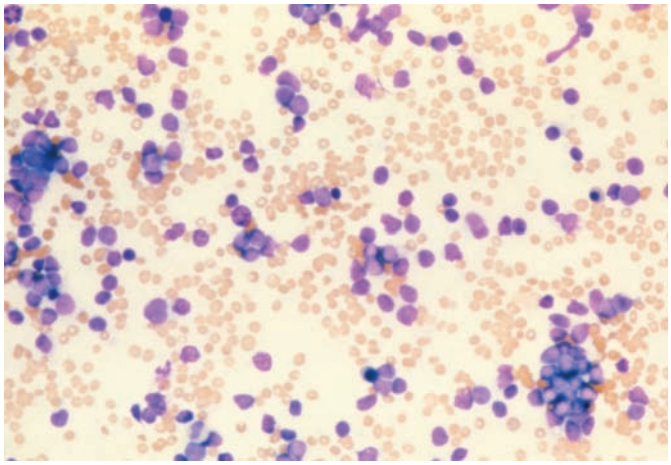


Figure 34–7 ■ A blood smear from the bone marrow of a client with acute lymphocytic leukemia. Note the abnormally large number of lymphocytes (stained purple) crowding the bone marrow. As a result, normal production of RBCs, functional WBCs, and platelets is suppressed.

Source: Dr. Gopal Murti, Photo Researchers, Inc.

Chronic Lymphocytic Leukemia

Chronic lymphocytic leukemia is characterized by proliferation and accumulation of small, abnormal, mature lymphocytes in the bone marrow, peripheral blood, and body tissues. The abnormal cells are usually B lymphocytes that are unable to produce adequate antibodies to maintain normal immune function. Only about 5% of CLL involves T cells (Copstead & Banasik, 2005). CLL occurs more commonly in adults, especially in older adults (median age 65). CLL is the least common type of the major leukemias.

CLL has a slow onset and is often diagnosed during a routine physical examination. If symptoms are present, they usually include vague complaints of weakness or malaise. Possible clinical findings include anemia, infection, and enlarged lymph nodes, spleen, and liver. As in other leukemias, bone marrow hyperplasia is present. Erythrocyte and platelet counts are reduced. Leukocyte counts may either be elevated or reduced, but abnormal cells are always present. In CLL, years may elapse before treatment is required. Survival of this disease averages approximately 7 years.

INTERDISCIPLINARY CARE

Treatment for leukemia focuses on achieving remission or cure and relieving symptoms. The methods of treatment may include chemotherapy, radiation therapy, and bone marrow or stem cell transplantation. Cure is more often achieved in children with acute leukemia than in adults, although long-term remissions (disease-free periods with no signs or symptoms) often can be achieved.

Diagnosis

The following diagnostic tests are ordered when leukemia is suspected:

- *CBC* with differential is done to evaluate cell counts, hemoglobin and hematocrit levels, and the number, distribution, and morphology (size and shape) of WBCs.

TABLE 34–5 Diagnostic Findings by Type of Leukemia

TEST	AML	CML	ALL	CLL
RBC count	Low	Low	Low	Low
Hemoglobin	Low	Low	Low	Low
Hematocrit	Low	Low	Low	Low
Platelet count	Very low	High early, low late	Low	Low
WBC count	Varies	Increased	Varies	Increased
Myeloblasts	Present			
Neutrophils	Decreased	Increased	Decreased	Normal
Lymphocytes		Normal		Increased
Monocytes		Normal/low		
Blasts	Present	Present (crisis)	Present	
Bone Marrow	Hypercellular		Hypercellular	
Myeloblasts	Present			
Lymphoblasts			Present	
Lymphocytes				Present

- *Platelets* are measured to identify possible thrombocytopenia secondary to the leukemia and the risk of bleeding.
- *Bone marrow examination* provides information about cells within the marrow, the type of erythropoiesis, and the maturity of erythropoietic and leukopoietic cells.

Table 34–5 outlines usual diagnostic test results in the various forms of leukemia.

Chemotherapy

Single agent or combination chemotherapy is the treatment of choice for most types of leukemia, with the goal of eradicating leukemic cells and producing remission. Table 34–6 outlines typical chemotherapy regimens for different types of leukemia. Combination chemotherapy reduces drug resistance and toxicity, and interrupts cell growth at various stages of the cell cycle, producing a complementary effect of the drugs used. Cancer treatment with chemotherapy is discussed in detail in Chapter 14 ∞.

Chemotherapy for leukemia generally is divided into the induction phase and postremission therapy. During *induction,* drug doses are high to eradicate leukemic cells from the bone marrow. These high doses often also damage stem cells and interfere with production of normal blood cells. Circulating mature blood cells are not affected because they are no longer dividing. The degree of bone marrow suppression is influenced by a number of factors, including age, nutritional status, concurrent chronic diseases such as impaired liver or renal function, the drug and drug dose, and prior treatment.

Colony-stimulating factors (CSFs), also called hematopoietic growth factors, often are administered to "rescue" the bone marrow following induction chemotherapy. CSFs are cytokines that regulate the growth and differentiation of blood cells. Factors that support neutrophil maturation, *granulocyte-macrophage CSF (GM-CSF)* and *granulocyte CSF (G-CSF*), are commonly used. Bone pain is a common side effect of therapy with these agents. Clients also may experience fevers, chills, anorexia, muscle aches, and lethargy (Kasper et al., 2005).

Once remission has been achieved, postremission chemotherapy is continued to eradicate any additional leukemic cells, prevent relapse, and prolong survival. A single chemotherapeutic agent, combination therapy, or bone marrow transplant may be used for postremission treatment.

Radiation Therapy

Radiation therapy damages cellular DNA. While the cell continues to function, it cannot divide and multiply. Cells that divide rapidly, such as bone marrow and cancer cells (radiosensitive cells), respond quickly to radiation therapy.

TABLE 34–6 Chemotherapeutic Regimens Used to Treat Leukemia

Acute myeloid leukemia	■ Cytarabine (Cytoxan, an alkylating agent), *with* daunorubicin (Cerubidine, an antitumor antibiotic) *or* idarubicin (Idamycin, an antitumor antibiotic)
	■ All-*trans* retinoic acid (ATRA) added for clients with promyelocytic leukemia
Chronic myeloid leukemia	■ Imatinib mesylate (Gleevec), a *bcr/abl* tyrosine kinase (enzyme) inhibitor
	■ Hydroxyurea (a DNA inhibitor) *or* homoharringtonine (HHT, a plant alkaloid) if imatinib not tolerated
Acute lymphocytic leukemia	■ Daunorubicin (Cerubidine, an antitumor antibiotic) *with* vincristine (Oncovin, a plant alkaloid) *with* prednisone *with* asparaginase (Elspar)
Chronic lymphocytic leukemia	■ Fludarabine (Fludara, an antimetabolite) *or* chlorambucil (Chloromycetin, an antitumor antibiotic)
	■ Cyclophosphamide (Cytoxan, an alkylating agent), vincristine, and prednisone
	■ Cyclophosphamide, doxorubicin (Adriamycin, an antitumor antibiotic), vincristine, and prednisone

Although normal cells are affected, they are better able to recover from the damage caused by the radiation than are cancer cells. The types of delivery, effects, and toxicities of radiation are discussed in greater detail in Chapter 14 ∞.

Bone Marrow Transplant

Bone marrow transplant (BMT) is the treatment of choice for some types of leukemia (see Table 34–3). BMT often is used in conjunction with or following chemotherapy or radiation. There are two major categories of BMT: In allogeneic BMT, the bone marrow of a healthy donor is infused into the client with the illness; in autologous BMT, the client is infused with his or her own bone marrow.

ALLOGENEIC BMT *Allogeneic BMT* uses bone marrow cells from a donor (often from a sibling with closely matched tissue antigens; closely matched unrelated donors also may be used). Prior to allogeneic BMT, high doses of chemotherapy and/or total body irradiation are used to destroy leukemic cells in the bone marrow. The donor's bone marrow is aspirated (Figure 34–8 ■) and infused through a central venous line into the recipient. Prior to BMT and reestablishment of bone marrow function, the client is critically ill and at significant risk for infection and bleeding due to depletion of WBCs and platelets.

AUTOLOGOUS BMT *Autologous BMT* uses the client's own bone marrow to restore bone marrow function after chemotherapy or radiation. This procedure is often called *bone marrow rescue*. In autologous BMT, about 1 L of bone marrow is aspirated (usually from the iliac crests) during a period of disease remission. The bone marrow is then frozen and stored for use after treatment. If relapse occurs, lethal doses of chemotherapy or radiation are given to destroy the immune system and malignant cells, and to prepare space in the bone marrow for new cells. The filtered bone marrow is then thawed and infused intravenously through a central line. The infused marrow cells slowly become a part of the client's bone marrow, the neutrophil count increases, and normal hematopoiesis takes place.

As in allogeneic BMT, the client is critically ill during the period of bone marrow destruction and immunosuppression. The client is hospitalized in a private room for 6 to 8 weeks or more. Potential complications include malnutrition, infection, and bleeding.

Figure 34–8 ■ Allogeneic bone marrow transplant. Bone marrow from the donor is aspirated, then filtered and infused into the recipient.

Source: Simon Fraser, Photo Researchers, Inc.

Stem Cell Transplant

Allogeneic **stem cell transplant (SCT)** is an alternative to bone marrow transplant. SCT results in complete and sustained replacement of the recipient's blood cell lines (WBCs, RBCs, and platelets) with cells derived from the donor stem cells.

Donors must have tissue that is closely matched with that of the recipient. Prior to harvesting, hematopoietic growth factors, including G-CSF and GM-CSF, are administered to the donor for 4 to 5 days. This increases the concentration of stem cells in peripheral blood, allowing it to be used for the transplant instead of bone marrow. Peripheral blood is removed and white cells are separated from the plasma, then administered via a large central venous catheter. Large concentrations of stem cells also are present in umbilical cord blood. This may be stored and used in some cases (Kasper et al., 2005).

The recipient undergoes similar treatment prior to SCT as for BMT. The risks for infection and other complications, as well as graft-versus-host disease, are similar.

Graft-Versus-Host Disease

Allogeneic BMT or SCT may precipitate *graft-versus-host disease (GVHD),* which develops in up to 60% of all clients receiving an allogeneic BMT or SCT (Kasper et al., 2005). In GVHD, immune cells of the donated bone marrow identify the recipient's body tissue as foreign. Consequently, T lymphocytes in the donated marrow attack the liver, skin, and GI tract, causing skin rashes progressing to desquamation (loss of skin), diarrhea, GI bleeding, and liver damage. *Acute GVHD* develops within days or weeks of the transplant and is usually marked by a pruritic, maculopapular rash that begins on the palms and soles of the feet, and may extend over the entire body. Vaso-occlusive disease of the liver affects up to 25% of allogeneic bone marrow transplant recipients, with jaundice and elevated liver function tests (Porth, 2005). *Chronic GVHD* develops later, 100 or more days after the transplant, affecting 20% to 50% of clients who survive 6 months or more following allogeneic BMT or SCT (Kasper et al., 2005). It may follow acute GVHD or develop in clients with no prior symptoms. GVHD is treated with antibiotics and steroids; immunosuppressant drugs such as thalidomide and immunotoxin (XomaZyme) may be used if necessary.

Biologic Therapy

Cytokines such as interferons and interleukins are biologic agents that may be used to treat some leukemias. These agents modify the body's response to cancer cells; in some cases they are cytotoxic as well. Interferons are a complex group of messenger pro-

teins normally produced in response to antigens such as viruses (see Chapter 12 ∞). They have multiple effects, including moderating immune function and inhibiting abnormal cell proliferation and growth. Interferon alpha may be used to treat some leukemias, particularly CML. Side effects commonly associated with interferon therapy include flulike symptoms, persistent fatigue and lethargy, weight loss, and muscle and joint pain.

Complementary Therapies

Although many complementary and alternative medicine therapies have been purported to treat cancer in general, at this time none have been shown to have sustained benefit in treating leukemia. Clinical trials have demonstrated the efficacy of both coping skills training (relaxation and imagery) and hypnosis to significantly reduce oral discomfort associated with leukemia and its treatment (Spencer & Jacobs, 2003).

NURSING CARE

For nursing care specific to the client undergoing diagnostic testing for leukemia, see the accompanying Nursing Care Plan.

Health Promotion

Health promotion activities related to leukemia include teaching about leukemia risk factors, particularly those that can be controlled. Discuss the potential dangers of exposure to ionizing

NURSING CARE PLAN A Client with Acute Myelocytic Leukemia

Catherine Cole is a 37-year-old secretary who lives with her husband, Ray, and teenage daughter, Amy, in an apartment in a large metropolitan area. About 2 months ago, Mrs. Cole began to tire easily and experience night sweats several times a week. She also noted that she was pale, bruised easily, and was having heavier menstrual periods. Blood tests ordered by her primary care provider are abnormal. She is admitted for a bone marrow biopsy.

ASSESSMENT

Mary Losapio, RN, obtains a nursing history and physical assessment for Mrs. Cole. Mrs. Cole tells her, "I'm so tired, and I have these bruises all over me. I'm so afraid of the results of the bone marrow examination. I don't know what we will do if I have cancer." Mrs. Cole clutches her husband's hand and then begins to cry. Physical assessment data include height, 64 inches (156 cm); weight, 106 lb (48.1 kg); vital signs: T 100°F, P 102, R 22, BP 130/82. Numerous petechiae scattered over trunk and arms; ecchymoses noted on lower right arm and right calf. Oral mucosa is red, with several small ulcerations in buccal areas.

Blood count shows reduced RBCs, hemoglobin, and hematocrit levels. The WBC is high, with myeloblasts seen on differential. The platelet count is very low. A tentative diagnosis of acute myelogenous leukemia is made.

DIAGNOSES

- *Risk for Infection* related to altered WBC production and immune function
- *Ineffective Protection* related to reduced platelet count and risk for bleeding
- *Impaired Oral Mucous Membrane* secondary to anemia and reduced platelets
- *Fatigue* related to anemia
- *Anxiety* related to fear of leukemia diagnosis

EXPECTED OUTCOMES

- Remain free of infection.
- Experience no significant bleeding.
- Have intact oral mucous membranes.
- Manage self-care activities despite fatigue.
- Verbalize decreased anxiety.

PLANNING AND IMPLEMENTATION

- Place in a private room.
- Limit visitors to immediate family for the present.
- Instruct all staff, the family, and client to carefully wash hands. Post a sign over the washbasin in the room as a reminder.
- Record vital signs every 4 hours.
- Avoid invasive procedures unless absolutely necessary.
- Monitor for bleeding every 4 hours, including skin, oral mucosa, abdominal assessment, body fluids, and menstrual pad count.
- Instruct to perform oral hygiene every 2 to 4 hours, using a soft-bristle toothbrush.
- Ask the dietitian to work with Mrs. Cole to identify preferred foods. Instruct to avoid foods that may damage oral mucosa, such as very hot, very cold, or highly acidic or spicy foods.
- Provide for periods of rest alternating with activity.
- Teach about the bone marrow biopsy. Allow time for questions and to verbalize fears.
- Refer to the oncology nurse specialist for further teaching and support.

EVALUATION

The bone marrow biopsy confirms the diagnosis of acute myelogenous leukemia. Mrs. Cole is very upset, but calms as the physician and the oncology nurse discuss treatment plans and the possibility of remission. She decides to have outpatient chemotherapy. During her hospital stay, Mrs. Cole remained free of infection or further bleeding. She tells Ms. Losapio that her mouth feels better, although it is still painful. During routine assessment, Mrs. Cole remarks, "You know, I was so scared when I came here, but I think I am a little less so now. Sometimes not knowing what is wrong is worse than knowing."

CRITICAL THINKING IN THE NURSING PROCESS

1. Describe how alterations in WBCs can increase a person's susceptibility to infection.
2. List sources of potential infection for the hospitalized client.
3. What is the rationale for having the client do her own oral and physical hygiene?
4. Outline a teaching plan for this client and her family for home care to prevent infection.
5. Develop a care plan for Mrs. Cole for the nursing diagnosis *Activity Intolerance.*

See Evaluating Your Response in Appendix C.

radiation and certain chemicals such as benzene. Encourage all clients to avoid smoking cigarettes. Discuss genetic counseling with clients at high risk for having a child with Down syndrome (over age 35).

Assessment

Focused assessment data related to leukemia include:

- *Health history:* Complaints of fatigue, weakness, dyspnea on exertion, frequent infections, sore throat, night sweats, bleeding gums, or nose bleeds; recent weight loss; exposure to ionizing radiation (multiple x-rays, residence near a site of radiation or atomic testing) or chemicals (occupational); prior treatment for cancer; history of an immune disorder.
- *Physical examination:* Skin and mucous membranes for bruising, purpura, petechiae, ulcers or lesions; pallor; vital signs including orthostatic vitals; heart and lung sounds; abdominal examination; stool for occult blood.
- *Diagnostic tests:* Blood count with differential; bone marrow studies.

Nursing Diagnoses and Interventions

When caring for the client with leukemia, the nurse considers the chronic and life-threatening nature of the disease as well as the effects of treatment. See the Nursing Research box below. Priority nursing problems may include *Risk for Infection, Imbalanced Nutrition: Less than Body Requirements, Impaired Oral Mucous Membranes, Ineffective Protection (Bleeding),* and *Anticipatory Grieving.*

MEDIALINK Care Plan Activity: Acute Myelocytic Leukemia

Risk for Infection

Changes in white blood cell function impair the immune and inflammatory responses in leukemia, increasing the risk for infection. WBCs may be immature and ineffective or, in some cases, deficient. Chemotherapy or radiation therapy further depresses bone marrow function and increases the risk for infection.

- Promptly report manifestations of infection: fever, chills, throat pain, cough, chest pain, burning on urination, purulent drainage, and itching and burning in vaginal or rectal areas. *Prompt reporting allows timely intervention to prevent overwhelming infection and sepsis.*
- Institute infection protection measures:
  a. Maintain protective isolation as indicated.
  b. Ensure meticulous hand washing among all people in contact with the client.
  c. Assist as needed with appropriate hygiene measures.
  d. Restrict visitors with colds, flu, or infections.
  e. Provide oral hygiene after every meal.
  f. Avoid invasive procedures when possible, including injections, intravenous catheters, catheterizations, and rectal and vaginal procedures. When necessary, use strict aseptic technique for all invasive procedures and monitor carefully for infection.

  These precautions minimize exposure to bacterial, viral, and fungal pathogens. Infection is the major cause of death in clients with leukemia. Mucous membranes are especially susceptible to breakdown and infection as a result of tissue damage from chemotherapy or radiation.

NURSING RESEARCH Evidence-Based Practice for Clients with Acute Leukemia and Lymphoma

Clients with acute leukemia and malignant lymphoma experience a number of distressing manifestations of their disease, including malaise and fatigue, fever, night sweats, infections, and possible hemorrhage. Treatments such as radiation therapy and chemotherapy often have numerous adverse effects as well, including anorexia and nausea, stomatitis, lethargy, malaise, and fatigue. In these studies, clients in remission from acute leukemia or malignant lymphoma were surveyed regarding physical problems, their view of help they received and who was of most help during treatment, and the impact of the disease and treatment on their current life (Persson, Hallberg, & Ohlsson, 1997; Persson & Hallberg, 2004).

Clients identified energy loss and nutritional problems as being most troublesome during disease treatment. In general, clients with more physical problems were less satisfied with the nursing care they received, suggesting that nurses were less effective in meeting the needs of the sickest clients. Clients continued to experience reduced psychologic and sexual energy and a significant need for intimate help and counseling during remission. While family relationships improved, work and finances were negatively impacted by their disease.

IMPLICATIONS FOR NURSING

This study points out the need for nurses to actively focus their care on the physical problems experienced during treatment, especially energy loss and nutritional problems. Overwhelming fatigue interferes with the client's ability to provide self-care, but its effects may not be readily apparent to nurses. The long-term effects of reduced psychologic and sexual energy, as well as continued susceptibility to infections, indicate a need for continued follow-up care, teaching, and possibly referral to counseling services.

CRITICAL THINKING IN CLIENT CARE

1. Explain the physiologic responses to malignancies and cancer treatments that cause fatigue, malaise, and nutritional problems.
2. Clients undergoing treatment for leukemia, malignant lymphoma, and other cancers may have few outward manifestations of their disease or responses to treatment. Discuss how this apparent well-being may affect the nurses' perception of care needs.
3. How may continued problems of fatigue and lack of psychologic and sexual energy affect family relations?
4. Develop a nursing care plan for a client with acute leukemia to address the nursing diagnosis *Ineffective Sexuality Patterns* related to fatigue and lack of energy.

- Monitor vital signs including temperature and oxygen saturation every 4 hours. Report temperature spikes with chilling, tachypnea, tachycardia, restlessness, change in Pao_2, and hypotension. *The inflammatory response may be impaired in leukemia, masking signs of infection until sepsis develops, indicated by manifestations such as those above.*
- Monitor neutrophil levels (measured in cubic millimeters) for relative risk for infection:
  2000 to 2500: no risk
  1000 to 2000: minimal risk
  500 to 1000: moderate risk
  Below 500: severe risk.
  Neutrophils are the first line of defense against infection. As levels decrease, the risk for infection increases.
- Explain infection precautions and restrictions and their rationale; explain that these measures are usually temporary. *Client and family understanding increases compliance and lowers the risk of infection.*

Imbalanced Nutrition: Less than Body Requirements

The client with leukemia may have difficulty meeting nutritional needs due to increased metabolism, fatigue, loss of appetite from radiation, nausea and vomiting from chemotherapy, or painful oral mucous membranes that make chewing and swallowing difficult and/or painful.

- Weigh regularly and evaluate weight loss over time to determine degree of malnutrition. A weight loss of 10% to 20% may indicate malnutrition. *A minimum intake of nutrients is necessary for health and tissue repair; cancer increases metabolic needs over this basal requirement. Weight loss occurs when metabolic requirements are not met. Both the disease process and its treatment can interfere with nutrient intake.*
- Address causative or contributing factors to inadequate food and fluid intake.
  a. Provide mouth care before and after meals; use a soft toothbrush or sponges as necessary.
  b. Provide liquids with different textures and tastes.
  c. Increase liquid intake with meals.
  d. Reduce intake of milk and milk products, which makes mucus more tenacious.
  e. Assist to a sitting position for eating.
  f. Ensure that the environment is clean and odor free.
  g. Provide medications for pain or nausea 30 minutes before meals, if prescribed.
  h. Provide rest periods before meals.
  i. Offer small, frequent meals including low-fat, high-kilocalorie foods throughout the day.
  j. Provide commercial supplements, such as Ensure.
  k. Avoid painful or unpleasant procedures immediately before or after meals.
  l. Suggest measures to improve food tolerance, such as eating dry foods when arising, consuming salty foods if allowed, and avoiding very sweet, rich, or greasy foods.

  Anorexia, nausea and vomiting, diarrhea, stomatitis, taste changes, and dysphagia often make eating difficult during cancer treatment when good nutrition is most important. Maintaining nutritional status decreases morbidity and mortality by preventing weight loss, improving the response to treatment, minimizing adverse effects, and improving quality of life. Small, frequent meals are often better tolerated, especially high-protein, high-kilocalarie foods.

Impaired Oral Mucous Membrane

Stomatitis, inflammation and ulceration of the oral mucous membrane, is common in leukemia. Chemotherapy can further impair the integrity of constantly dividing oral tissues.

- Inspect the buccal region, gums, sublingual area, and the throat daily for swelling or lesions. Ask about oral pain or burning. *Breakdown of the oral mucous membrane increases the risk of infection and bleeding, causes pain and discomfort with eating and swallowing, and may cause swelling that interferes with the airway.*
- Culture any oral lesions. *Herpes simplex virus and* Candida *(yeast) are more common in clients with neutropenia. Herpes lesions are usually red, raised, fluid-filled blisters;* Candida *causes a white coating and patches of white plaque.*
- Assist with mouth care and oral rinses with saline or a solution of hydrogen peroxide and water (1:1 or 1:3 hydrogen peroxide and water) every 2 to 4 hours. Apply petroleum jelly to the lips to prevent dryness and cracking. *These measures help prevent infection and increase comfort.*
- Encourage use of soft-bristle toothbrush or sponge to clean teeth and gums. *Toothbrushes with hard bristles may abrade inflamed mucosa, causing bleeding and increasing the risk of infection.*
- Administer medications as ordered to treat infection or relieve pain. *Topical antifungal agents such as nystatin may be prescribed to treat* Candida *infections. Topical anesthetics such as lidocaine may be prescribed to relieve comfort and facilitate good oral care.*
- Instruct to avoid alcohol-based mouthwashes, citrus fruit juices, spicy foods, very hot or very cold foods, alcohol, and crusty foods. Suggest bland, cool foods and cool liquids at least every 2 hours. *Avoiding mucosa-traumatizing foods and liquids increases comfort; bland, cool foods and liquids cause the least pain. Intake of adequate fluids is necessary to prevent dehydration.*

Ineffective Protection

Bleeding is the second most common cause of leukemia deaths. As platelet counts decrease, the risk of bleeding increases (see the section later in this chapter on thrombocytopenia). Tumor lysis syndrome also is a risk in clients with leukemia who are undergoing their initial treatment with chemotherapy. Tumor lysis syndrome develops when a large number of malignant cells are destroyed by treatment with chemotherapy or radiation. The resultant by-products of cell lysis can overwhelm the body's ability to effectively eliminate them, leading to hyperkalemia, hyperphosphatemia with secondary hypocalcemia, and hyperuricemia (Cantril & Haylock, 2004).

- Assess vital signs every 4 hours and body systems every shift for bleeding:
  a. Skin and mucous membranes for petechiae, ecchymoses, and purpura

b. Gums, nasal membranes, and conjunctiva for bleeding
c. Vomitus, stool, and urine for visible or occult blood
d. Vaginal bleeding
e. Prolonged bleeding from puncture sites
f. Neurologic changes such as headache, visual changes, altered mentation, decreased level of consciousness, seizures
g. Abdomen for complaints of epigastric pain, diminished bowel sounds, increasing abdominal girth, rigidity or guarding.

Early identification of bleeding helps prevent significant blood loss and potential shock. Internal hemorrhage may lead to tachycardia, hypotension, pallor, and diaphoresis. Bleeding into the lungs may cause dyspnea; bleeding into the abdomen causes increased girth, pain, and guarding. Intracranial bleeding affects mental status and level of consciousness.

- Avoid invasive procedures such as rectal temperatures and suppositories, vaginal douches, suppositories, tampons, urinary catheterization, and parenteral injections if possible. Diagnostic procedures such as biopsy or lumbar puncture should not be done if the platelet count is less than 50,000. *Invasive procedures can cause tissue trauma and bleeding. Procedures that use large-bore needles should be delayed until the platelet count is increased.*
- Apply pressure to injection sites for 3 to 5 minutes, and to arterial punctures for 15 to 20 minutes. *Pressure prevents prolonged bleeding by prompting hemostasis and clot formation.*
- Instruct to avoid forcefully blowing or picking the nose, forceful coughing or sneezing, and straining to have a bowel movement. *These activities can damage mucous membranes, increasing the risk for bleeding.*
- Monitor and promptly report abnormal blood levels of electrolytes, uric acid, urea nitrogen, and creatinine, or manifestations of tumor lysis syndrome. *Significant alterations in electrolyte levels can lead to complications such as cardiac dysrhythmias, muscle weakness or tetany, paresthesias, and mental status changes. Excess uric acid can compromise renal function, and lead to metabolic acidosis and gout.*
- Maintain adequate hydration and administer prescribed medications such as allopurinol and diuretics as ordered. *Hydration is vital to maintain renal function and promote elimination of tumor lysis by-products. Allopurinol reduces the risk of uric acid crystallization in the kidneys and other tissues* (Cantril & Haylock, 2004).

Anticipatory Grieving

The diagnosis of cancer and a potentially life-threatening illness causes actual or perceived losses, such as loss of function, independence, normal appearance, friends, self-esteem, and self. Grieving is the emotional response to those losses. The adaptive process of mourning a loss and resolving grief is called grief work; grief work cannot begin until a loss is acknowledged. See Chapter 5 ∞ for a detailed discussion of grief and loss.

- Discuss roles of the client and family and ways in which they managed stressful situations in the past. Assess coping strategies and their effectiveness. Help identify sources of strength and support. Discuss changing roles resulting from leukemia diagnosis, and its effect on spiritual, social, and economic status and usual lifestyle. Evaluate cultural or ethnic factors that affect grief reactions. *Grieving is a normal response to a real or potential loss that begins at the time of diagnosis. The timing, duration, and intensity of grief and responses to grief may differ among family members. Share information on diagnosis, role change, and physical loss among all family members to build the foundation for mutual understanding and trust.*
- Use therapeutic communication skills to facilitate open discussion of losses and provide permission to grieve. *Encouraging discussion of the meaning of the loss helps decrease some of the anxiety associated with loss. This in turn allows the client and family to examine the current situation and compare it with past situations that they have coped with successfully.*
- Provide information about agencies that may help in resolving grief, and make referrals as indicated. Consider self-help groups, cancer support groups, and bereavement groups. *Participating in support groups with others who are anticipating or experiencing a similar loss can decrease feelings of isolation.*

Linking NANDA, NIC, and NOC

Chart 34–2 shows links between NANDA nursing diagnoses, NIC, and NOC for the client with leukemia.

NANDA, NIC, AND NOC LINKAGES
CHART 34–2 **The Client with Leukemia**

NANDA

Ineffective Protection

↓

NIC

Bleeding Precautions
Energy Management
Infection Protection
Neurologic Monitoring
Oral Health Restoration

↓

NOC

Coagulation Status
Energy Conservation
Immune Status
Neurological Status: Consciousness
Tissue Integrity: Skin and Mucous Membranes

Data from *NANDA's Nursing Diagnoses: Definitions & Classification 2005–2006* by NANDA International (2005), Philadelphia; *Nursing Interventions Classification (NIC)* (4th ed.) by J. M. Dochterman & G. M. Bulechek (2004), St. Louis, MO: Mosby; and *Nursing Outcomes Classification (NOC)* (3rd ed.) by S. Moorhead, M. Johnson, and M. Mass (2004), St. Louis, MO: Mosby.

Community-Based Care

Client and family teaching for home care after treatment for leukemia focuses on encouraging self-care, providing information about the disease and the treatment, preventing infection and injury, and promoting nutrition. Teaching topics for each of these areas are as follows.

Encouraging Self-Care

- Hygiene measures and energy conservation during self-care activities
- Oral hygiene including using a soft-bristle toothbrush several times daily; avoiding flossing
- Reporting lesions, bleeding, or signs of infection promptly
- Maintaining a balance of rest and activity.

Information about Leukemia and Treatment

- Bone marrow function, the pathophysiology of leukemia, and potential complications of leukemia
- Prognosis for the specific type of leukemia
- Treatment measures such as chemotherapy, radiation, bone marrow or stem cell transplant, their purpose and effects, where treatment is available, and potential adverse effects or risks
- Community, regional, and national resources for people with leukemia.

Preventing Infection and Injury

- Hand washing and other measures to reduce exposure to pathogens such as avoiding people who are ill and avoiding crowds
- Avoiding foodborne illnesses by washing fruits and vegetables, proper food storage
- Dental hygiene measures
- Avoiding immunizations
- Manifestations to report: fever, chills, burning on urination, foul-smelling urine, vaginal or rectal discharge, skin lesions
- Avoiding contact sports or strenuous exercise if platelet count is low
- Using an electric razor for shaving, avoiding rectal or vaginal suppositories, vaginal tampons, or enemas
- Increasing dietary fiber and using a bulk-forming laxative as needed to prevent straining
- Avoiding over-the-counter or prescription drugs that interfere with platelet function (see Box 34–6 on page 1140)
- The importance of reporting any bleeding (nosebleeds, rectal bleeding, vomiting blood, excessive menstrual periods, blood in the urine, bleeding gums, bruises, or collections of blood under the skin) or changes in behavior to the healthcare provider.

Promoting Nutrition

- Eating several small, low-fat, high-calorie meals and drinking five to eight glasses of water daily
- Reporting continued weight loss, loss of appetite, or inability to eat for 24 hours
- Discussing dietary needs with the dietitian.

Assistance with physical care, finances, and transportation may be required following discharge. Refer the client and family to social services, support groups, home care services as needed, and other agencies that can provide needed services (such as local chapters of the American Cancer Society, which can provide hospital beds and transportation for outpatient cancer treatment).

THE CLIENT WITH MALIGNANT LYMPHOMA

Lymphomas are malignancies of lymphoid tissue. They are characterized by the proliferation of lymphocytes, histiocytes (resident monocytes or macrophages), and their precursors or derivatives. Lymphomas are closely related to lymphocytic leukemias. Some experts consider them to be different forms or stages of the same disease processes.

Although there are many types of malignant lymphoid cells, at this time lymphomas commonly are identified as Hodgkin's disease or non-Hodgkin's lymphoma.

Incidence and Risk Factors

Malignant lymphomas are the seventh leading cause of cancer deaths in the United States. Approximately 63,740 new cases of lymphoma were diagnosed in 2005, and 20,600 deaths were attributed to the disease. The incidence of non-Hodgkin's lymphoma has nearly doubled since 1970, but currently has stabilized, primarily due to a fall in its incidence related to HIV infection and AIDS. The incidence of Hodgkin's disease has significantly declined since 1990 (ACS, 2005).

While the cause of lymphoma is unknown, some risk factors have been identified. See the accompanying Genetics Considerations box for information about identified genetic links for lymphoma development. Immunosuppression due to drug therapy following organ transplant or to HIV disease increases the risk for non-Hodgkin's lymphoma. Infectious agents such as HTLV-1 and EBV also have been identified as risk factors. Others may include occupational herbicide or chemical exposure (ACS, 2005).

Pathophysiology

Hodgkin's Disease

Hodgkin's disease is a lymphatic cancer, occurring most often in people between the ages of 15 and 35 or over age 50. It is somewhat more common in men than women. Approximately

GENETIC CONSIDERATIONS
Focus on Lymphoma

Although specific genetic alterations have not been identified for all types of lymphoma, recurring genetic abnormalities associated with lymphomas point to a genetic link in disease development. Three distinct genetic abnormalities have been identified in non-Hodgkin's lymphomas: gross chromosomal changes such as translocations, rearrangements of specific genes, and altered expression of specific oncogenes (overexpression, underexpression, or mutation). Consistent genetic changes are associated with some lymphomas; in other cases, several genetic abnormalities may be seen. Hodgkin's disease is unique from other lymphomas in that no specific genetic abnormalities have been identified (Kasper et al., 2005). For more information about genetics and disease, see Chapter 8 ∞.

7350 new cases of Hodgkin's disease were diagnosed in 2005 (ACS, 2005). The exact cause of Hodgkin's disease is unknown, but both Epstein-Barr virus (EBV) infection and genetic factors appear to play a role in its development. Hodgkin's disease is one of the most curable cancers. As many as 60% to 90% of people with localized disease achieve cure with a normal life span (Porth, 2005).

Hodgkin's disease develops in a single lymph node or chain of nodes, spreading to adjoining nodes. Involved lymph nodes contain *Reed-Sternberg cells* (malignant cells) surrounded by host inflammatory cells. These malignant cells secrete inflammatory mediator substances, attracting inflammatory cells to the tumor site. They may invade almost any tissue in the body. The spleen often is involved; as the disease progresses, the liver, lungs, digestive tract, and CNS may be affected (Porth, 2005). Rapid proliferation of abnormal lymphocytes impairs the immune response, especially cell-mediated immune responses. Infections are common.

Hodgkin's disease is classified as classic Hodgkin's disease or as nodular lymphocyte-predominant Hodgkin's disease. The classic form of the disease accounts for 95% of all cases; nodular lymphocyte-predominant Hodgkin's is rare. Classic Hodgkin's can be further divided into four subtypes by cells identified within the tumor, but the subtype does not affect the prognosis (Copstead & Banasik, 2005; Kasper et al., 2005).

MANIFESTATIONS The most common symptom of Hodgkin's disease is one or more painlessly enlarged lymph nodes, usually in the cervical or subclavicular region. Systemic manifestations such as persistent fever, night sweats, fatigue, and weight loss are associated with a poorer prognosis for the disease. Late symptoms such as malaise, pruritus, and anemia indicate spread of the disease (Porth, 2005). The spleen may be enlarged, and other organ systems such as the lungs and gastrointestinal tract are occasionally involved.

Non-Hodgkin's Lymphoma

Non-Hodgkin's lymphoma is a diverse group of lymphoid tissue malignancies that do not contain Reed-Sternberg cells. Non-Hodgkin's lymphomas tend to arise in peripheral lymph nodes and spread early to tissues throughout the body. Non-Hodgkin's lymphoma is more common than Hodgkin's disease, affecting an estimated 56,390 people annually and causing about 19,200 deaths in 2005 (ACS, 2005). Older adults are more often affected, and it occurs more frequently in men than in women. Like Hodgkin's disease, its cause is unknown, although both genetic and environmental factors (e.g., viral infections such as EBV, human T-cell leukemia/lymphoma virus-1 [HTLV-1] and HTLV-2, and HIV) are thought to play a role.

As in most malignancies, non-Hodgkin's lymphoma begins as a single transformed cell; it may arise from T cells, B cells, or tissue macrophages (histocytes). The primary types of non-Hodgkin's lymphoma are identified in Table 34–7. Although non-Hodgkin's lymphoma usually arises in a lymph node, it can originate in any lymphoid tissue. It tends to spread early and unpredictably to other lymphoid tissues and organs. Extra-

TABLE 34–7 **Subtypes of Non-Hodgkin's Lymphoma**

SUBTYPE	INCIDENCE	COURSE AND PROGNOSIS
B-Cell Lymphomas		
Diffuse large B-cell lymphomas	Most common adult type (40%–50% of adult lymphomas) More common in males Incidence increases with aging	Aggressive tumor 45%–50% cure rate
Follicular lymphoma	Accounts for 40% of adult lymphomas, rare in children Incidence increases with aging	Bone marrow frequently involved Course slow, indolent; 72% 5-year survival
Extranodal marginal zone lymphoma (MALT lymphoma)	Accounts for about 5% of adult lymphomas, rare in children Incidence increases with aging More common in Italy	Presents with tumors outside lymphatic system: GI tract, lung, thyroid, urinary tract, skin, CNS Slow, indolent course; 74% 5-year survival
Mantle cell lymphoma	Accounts for 3% to 4% of adult lymphomas, rare in children Predominantly affects older men (74%)	Aggressive, difficult to cure 27% 5-year survival
Burkitt lymphoma	Rare in adults (<1% of lymphomas), more common in children (~30% NHL)	Rapidly progressive but responds well to therapy 45% 5-year survival
T-Cell Lymphomas		
Precursor T-cell lymphoblastic leukemia/lymphoma	More common in children and young adults More common in males than females	Can present either as ALL or lymphoma Aggressive disease; 26% 5-year survival
Peripheral T-cell lymphoma	Most common T-cell lymphoma in adults	Often presents as disseminated disease 25% 5-year survival
Mycosis fungoides/cutaneous T-cell lymphoma	Onset typically during mid-50s; more common in African Americans	Cutaneous lymphoma Slow course, progressing from patchy skin lesions to plaque to cutaneous tumors

nodal spread may involve the nasopharynx, gastrointestinal tract, bone, CNS, thyroid, testes, and soft tissue.

The prognosis for non-Hodgkin's lymphoma ranges from excellent to poor, depending on the identified cell type and grade of differentiation. Low-grade tumors (better differentiated) tend to be less aggressive and more curable. Higher grade tumors often are disseminated at the time of diagnoses, and have a poorer prognosis (Copstead & Banasik, 2005).

MANIFESTATIONS The early manifestations of non-Hodgkin's lymphoma are similar to those for Hodgkin's disease. Painless lymphadenopathy may be localized or widespread (Figure 34–9 ■). Systemic manifestations such as fever, night sweats, fatigue, and weight loss may be present, but are less common in non-Hodgkin's lymphoma. Organ system involvement may cause symptoms such as abdominal pain, nausea, and vomiting. Headaches, peripheral or cranial nerve symptoms, altered mental status, or seizures may signal CNS involvement.

The manifestations and clinical features of Hodgkin's disease and non-Hodgkin's lymphoma are compared in Table 34–8.

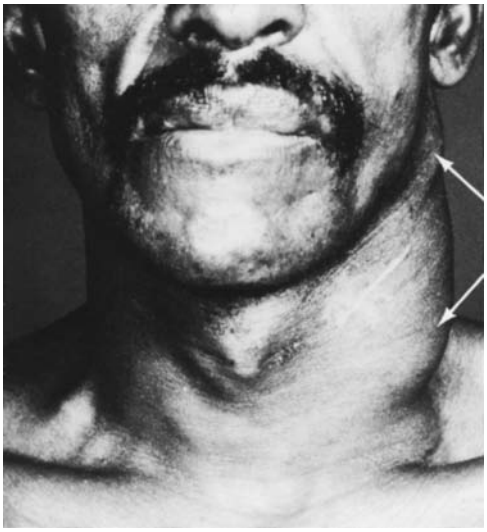


Figure 34–9 ■ Cervical lymphadenopathy in a client with lymphoma of the neck.

Source: Centers for Disease Control and Prevention (CDC)

Course

In both Hodgkin's disease and non-Hodgkin's lymphoma, the stage of the disease, the presence of systemic manifestations, and factors such as age help determine the prognosis. The prognosis is good when the disease is localized to one or two node regions. Factors such as anemia, thrombocytopenia, and older age reduce the likelihood of disease cure.

INTERDISCIPLINARY CARE

Chemotherapy and radiation therapy, either alone or in combination, are the primary treatments for Hodgkin's and non-Hodgkin's lymphomas. Use of monoclonal antibodies to target lymphoma cells, and bone marrow and peripheral stem cell transplants are under investigation for treating lymphomas as well. See the previous section on treatment of leukemia for more information about these transplants.

Diagnosis

The following diagnostic tests may be ordered for lymphomas:

- *CBC* often shows a mild normochromic, normocytic anemia in Hodgkin's disease; other findings in Hodgkin's disease may include leukocytosis with high neutrophil and eosinophil counts, and an elevated erythrocyte sedimentation rate (ESR). In non-Hodgkin's lymphoma, the CBC typically remains normal until late in the disease, when pancytopenia may develop.
- An ESR is done to identify possible inflammatory causes of lymph node enlargement.
- *Chemistry studies* of major organ function (including liver function tests and renal function studies) are performed to identify possible organ involvement. *Serum LDH levels* and *protein electrophoresis* also may be done when Hodgkin's disease is suspected.
- *Chest x-ray* is done to identify possible enlarged mediastinal lymph nodes and pulmonary involvement.
- *CT scans* of the chest, abdomen, and pelvis are performed to identify abnormal or enlarged nodes.
- *Positron emission tomography (PET or gallium scans)* may be performed in diagnosing the disease, as well as to evaluate the effectiveness of treatment.
- *Biopsy* of the largest, most central enlarged lymph node and of the bone marrow is done to establish the diagnosis for both

TABLE 34–8 **Features and Manifestations of Hodgkin's Disease and Non-Hodgkin's Lymphoma**

FEATURE OR MANIFESTATION	HODGKIN'S DISEASE	NON-HODGKIN'S LYMPHOMA
Lymphadenopathy	Localized to a single node or chain, often cervical, subclavicular, or mediastinal	Multiple peripheral nodes, nodes of the mesentery often involved
Spread	Orderly and continuous	Diffuse and unpredictable
Extranodal involvement	Rare	Early and common
Bone marrow involvement	Uncommon	Common
Fever, night sweats, weight loss	Common	Uncommon until disease is extensive
Other manifestations	Fatigue, pruritus, splenomegaly; anemia, neutrophilia	Abdominal pain, nausea, vomiting; dyspnea, cough; CNS symptoms; lymphocytopenia

Hodgkin's disease and non-Hodgkin's lymphoma. The presence of Reed-Sternberg cells confirms the diagnosis of Hodgkin's disease.

Staging

Staging is used to determine the extent of the disease and appropriate treatment. The Ann Arbor Staging System is used to assess the extent and severity of lymphomas. The stages are:

Stage I: involvement of a single lymph node region or lymphoid structure (e.g., spleen, thymus, lymphoid tonsillar tissue)

Stage II: involvement of two or more lymph node regions on the same side of the diaphragm

Stage III: involvement of lymph node regions or structures on both sides of the diaphragm

- III_1: limited to upper abdomen (spleen, splenic, celiac, or portal nodes)
- III_2: involvement of lower abdominal nodes (para-aortic, iliac, or mesenteric)

Stage IV: involvement of an extranodal site (not proximal or contiguous with an involved node) such as the liver, lung or pleura, bone or bone marrow, or skin.

The presence or absence of systemic symptoms is indicated by either an A (no systemic symptoms) or B (systemic symptoms of fever, night sweats, weight loss).

Chemotherapy

Combination chemotherapy is used to treat both Hodgkin's disease and non-Hodgkin's lymphoma. In both cases, chemotherapy often is followed by radiation therapy to involved lymph node regions. The choice of drug combination depends on the stage of the disease as well as the client's age and general condition. Combination regimens used in the United States include CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone); ABVD (doxorubicin, bleomycin, vinblastine, and dacarbazine); MOPP (nitrogen mustard, vincristine, procarbazine, and prednisone); and ChlVPP (chlorambucil, vinblastine, procarbazine, and prednisone). These regimens also may be combined in alternating months to reduce the adverse effects and improve tumor cell kill. More than 75% of clients with Hodgkin's disease who do not have systemic symptoms achieve complete remission with treatment. The prognosis for clients with non-Hodgkin's lymphoma varies by the type and stage of the disease. For more information about nursing care of the client receiving combination chemotherapy, see Chapter 14 ∞.

Radiation Therapy

Radiation therapy may be the primary treatment for early-stage Hodgkin's disease, although early chemotherapy is becoming more common. In later stages and in non-Hodgkin's lymphoma it usually is combined with chemotherapy. Many lymphomas are highly responsive to radiation. The involved lymph node region is treated, with careful shielding to protect unaffected areas and minimize the extent of radiation burn and normal cell destruction (Figure 34–10 ■). If the disease is advanced, total nodal irradiation may be done. See Chapter 14 ∞ for nursing care of the client receiving radiation therapy.

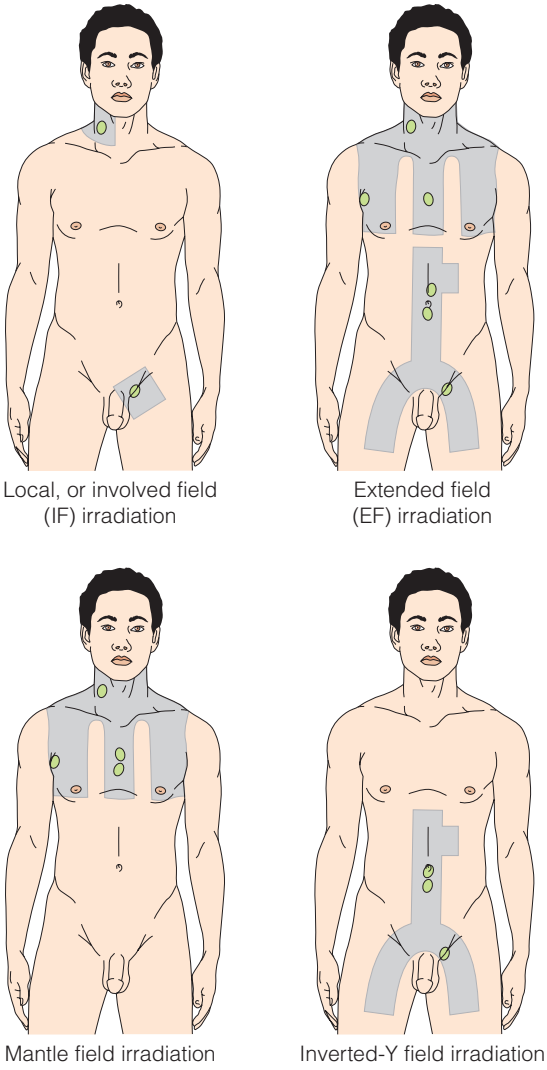


Figure 34–10 ■ Patterns of radiation therapy used to treat lymphoma based on the location and extent of the disease.

Stem Cell Transplant

Autologous peripheral blood stem cell transplant (PBSCT) is a treatment option for clients who experience remission of malignant lymphoma. Autologous PBSCT uses the client's own stem cells to restore bone marrow function after chemotherapy or radiation. In autologous PBSCT, stem cells are obtained from peripheral blood following chemotherapy and treatment with colony-stimulating factors to promote development of normal blood cells. The blood containing these normal stem cells is then frozen and stored for use after treatment. If relapse occurs, lethal doses of chemotherapy or radiation are given to destroy the immune system and malignant cells. The blood is then thawed and infused intravenously through a peripheral line. The infused stem cells become a part of the client's bone marrow and normal hematopoiesis takes place.

The client is critically ill during the period of bone marrow destruction and immunosuppression. The client is hospitalized in a private room for 6 to 8 weeks or more. See the accompanying nursing research box for discussion of fatigue related to autologous PBSCT.

NURSING RESEARCH Evidence-Based Practice: Client Undergoing Stem Cell Transplant

Fatigue and depression are common adverse effects of chemotherapy and radiation therapy in clients undergoing cancer treatment. Fatigue is prevalent, affecting 80% to 100% of people undergoing standard chemotherapy and often leading to lost work time and difficulty maintaining functional roles within the family and society. A study by El-Banna and colleagues (2004) sought to describe patterns of fatigue and depression and their relationship to one another among clients undergoing autologous PBSCT. Three time periods during therapy were selected for evaluation: prior to the initiation of chemotherapy, during chemotherapy just prior to transplant, and following the transplant. The setting for this study allowed transplant clients to reside in a hotel-like suite with a designated caregiver (family member or friend).

This study revealed that, among study participants, total fatigue and all its identified dimensions (behavioral, cognitive/mood, sensory, and affective) increased sharply at the time of the transplant, peaking at 7 days post-transplant. Depression followed the same pattern as fatigue, peaking at post-transplant day 7. Peak depression scores were indicative of major depression using the tool employed in this study.

IMPLICATIONS FOR NURSING

This study suggests that nurses should assess for fatigue and depression in clients following stem cell transplant. Nursing measures to help conserve the client's energy are appropriate to manage fatigue. Early detection and intervention for fatigue may actually reduce the intensity of fatigue at its peak. Nurses can use this information to prepare clients for common symptom patterns following SCT, thus reducing anxiety and concern that their condition may be declining rather than improving after SCT. Additionally, assessment tools to measure fatigue and depression among clients undergoing SCT should be incorporated into nursing care.

CRITICAL THINKING IN CLIENT CARE

1. The average age of participants ($N=27$) in this study was 49 years (range: 19 to 71 years); 56% were male, 44% female. Does this sample reflect the demographic of clients with malignant lymphoma and those undergoing autologous PBSCT? How can information about the sample be used to guide application of study findings to the general population of clients undergoing this treatment? To older adults? To adolescents and young adults?
2. In this study, participants resided in a hotel-like setting with a family member or friend who participated in care. How might you expect the results to differ had the clients been unable to closely interact with a caregiver of their own choosing?
3. Can the results of this study be applied when caring for clients undergoing allogeneic BMT or SCT? Why or why not?

Complications of Treatment

Both chemotherapy and radiation therapy may have long-term effects. Permanent sterility is common, especially in older adults. Bone marrow depression can lead to immunosuppression, anemia, and bleeding. Secondary cancers and cardiac injury are the most serious late adverse effects of treatment. Chemotherapy regimens using the MOPP or a related protocol carry a risk of acute leukemia. Cancers such as breast or lung cancer may develop 10 or more years after thoracic radiation. Thoracic radiation also increases the risk for coronary heart disease and hypothyroidism (Kasper et al., 2005).

NURSING CARE

Assessment and priority nursing care for the client with lymphoma follows. See also the Nursing Care Plan at the end of this section for application of nursing care strategies for a specific client with Hodgkin's disease.

Assessment

Focused assessment of the client with Hodgkin's disease or non-Hodgkin's lymphoma includes:

- *Health history:* Complaints of enlarged lymph node(s), fever, night sweats, weight loss, fatigue or general malaise, abdominal pain, respiratory symptoms, numbness or tingling of extremities, visual changes, or changes in mentation; history of infectious mononucleosis, HIV disease, or other immunosuppressive disorders.
- *Physical examination:* Mental status exam; inspect and palpate lymph nodes (cervical, subclavicular, axillary, and inguinal) for enlargement, tenderness; heart and lung sounds; abdominal examination for tenderness, masses, liver or spleen enlargement.
- *Diagnostic tests:* CBC, hemoglobin and hematocrit, ESR; serum chemistry results; x-ray, scan, and biopsy results.

Nursing Diagnoses and Interventions

Nursing care of the client with malignant lymphoma involves both physical and emotional support during diagnosis and treatment. Common nursing care problems include impaired protection due to bone marrow suppression, fatigue, nausea, and altered body image. See the nursing care section for leukemia for specific nursing interventions for *Ineffective Protection.*

Fatigue

General malaise and fatigue may accompany malignant lymphoma and are side effects of chemotherapy. In addition, the physical and psychologic stress of dealing with a chronic, debilitating disease and its treatment may cause fatigue.

- Inquire about feelings of malaise (a vague feeling of body weakness or discomfort) and fatigue (a pervasive, drained feeling that cannot be eliminated). *Both malaise and fatigue are subjective experiences with physiologic, situational, and psychologic components.*
- Encourage verbalization of feelings about the impact of the disease and fatigue on lifestyle. *Discussion of feelings helps the client clarify values and may assist in identifying priorities.*
- Encourage enjoyable but quiet activities, such as reading, listening to music, or hobbies. *Enjoyable activities help decrease feelings of fatigue. Quiet activities conserve energy while yielding a sense of accomplishment.*

NURSING CARE PLAN A Client with Hodgkin's Disease

Albin Quito, age 28, is the nurse manager of a thoracic intensive care unit in a large teaching hospital. Lately he has been more tired than usual, often wakes up at night covered with sweat, and just does not feel well. He had thought that his symptoms were due to a viral illness and his busy work schedule. However, yesterday morning Albin noticed a large swollen area on the right side of his neck. He made an appointment with his primary health provider who found a large cervical lymph node. A biopsy of the node and a CT scan of the chest were scheduled.

ASSESSMENT

David Herzog, the nurse in charge of the outpatient clinic, obtains a nursing history and assessment on Mr. Quito. His physical examination is essentially normal, with the exception of the enlarged node, which is not tender to palpation. When Mr. Quito is weighed, he tells Mr. Herzog that he has lost 7 lb (3.2 kg) in the past 2 months. In reviewing the results of the blood studies, Mr. Herzog notes mild anemia and an increased neutrophil count. The lymph node biopsy shows Reed-Sternberg cells. The clinic physician and Mr. Herzog tell Mr. Quito that the findings indicate stage 1-B Hodgkin's disease but that the prognosis is very good. The physician recommends a short course of combination chemotherapy followed by radiation therapy to involved sites.

DIAGNOSES

- *Anxiety* related to the diagnosis of Hodgkin's disease and effects of treatment on job performance
- *Risk for Infection* related to potential bone marrow depression due to chemotherapy
- *Fatigue* related to effects of cancer, chemotherapy, and radiation therapy

EXPECTED OUTCOMES

- Verbalize reduced anxiety.
- Remain free of infection.
- Identify and use methods to preserve energy.

PLANNING AND IMPLEMENTATION

- Encourage to consider a leave of absence from work during course of treatment.
- Discuss joining a support group for people with cancer.
- Provide information about the illness, combination chemotherapy, and radiation therapy.
- Reinforce knowledge of actions to decrease the risk of infection.
- Discuss ways to decrease fatigue and maintain energy:
  - Take a 1- to 2-hour nap once or twice a day.
  - Avoid overexertion during weekends and time off.
  - Maintain a well-balanced diet.

EVALUATION

When Mr. Quito returns the following week to begin chemotherapy, he brings his friend Nancy to meet Mr. Herzog and asks him to discuss his treatment with her. Mr. Quito says, "I am still really scared, but being able to talk about this with Nancy will help a lot." Mr. Quito has made arrangements to take a 4-month leave from work, with the understanding that his job will be held for him. He states that he will have some problems with money but is working them out. He also says he feels that taking a nap is silly but that he will rest to maintain his energy level. Mr. Quito and Nancy express confidence that he will be cured and say they plan to be active members of the cancer support group—even after recovery.

CRITICAL THINKING IN THE NURSING PROCESS

1. Discuss the rationale for treating Hodgkin's disease with chemotherapy and radiation.
2. Design a teaching plan to help Mr. Quito prevent infection while he is at home.
3. What effect does the diagnosis of cancer have on the developmental tasks of a young adult?
4. Develop a care plan for Mr. Quito for the nursing diagnosis *Ineffective Role Performance.*

See Evaluating Your Response in Appendix C.

- Encourage to establish priorities and include rest periods or naps when scheduling daily activities. *This provides a sense of control over activities and helps maintain self-esteem. Scheduled rest periods help restore energy and decrease fatigue.*
- Encourage delegation of some responsibilities to family members. *Delegation helps maintain the client's involvement and role in family decisions and responsibilities, while conserving energy for those activities identified as high priority by the client.*
- Identify and encourage the client to use energy-saving equipment. *Performing tasks with less exertion and in less time helps conserve energy.*
- Encourage a diet high in carbohydrates and fluids. *A high-carbohydrate diet helps maintain muscle glycogen stores. A liberal fluid intake promotes excretion of metabolic by-products that may contribute to malaise and fatigue.*

Nausea

The effects of malignant lymphoma and its treatment with chemotherapy and/or radiation therapy can contribute to nausea and interfere with nutritional status. Nausea, a sensation of abdominal fullness, and fear of vomiting often limit food intake. See also the nursing diagnosis *Imbalanced Nutrition* in the section on leukemia for additional interventions.

- Assess precipitating factors for nausea and/or vomiting, the frequency of vomiting, and relief measures used by the client. *Careful assessment allows development of interventions tailored to the client's situation and needs.*

PRACTICE ALERT

Provide ordered antiemetics before chemotherapy is started. Administering prescribed antiemetics before chemotherapy helps prevent nausea and the psychologic association of nausea with chemotherapy.

- Teach measures to prevent or relieve nausea and vomiting.
  a. Eat soda crackers and suck on hard candy.
  b. Eat soft, bland foods that are cold or at room temperature.
  c. Avoid unpleasant odors, and get fresh air.
  d. Eat prior to but not immediately before chemotherapy.

e. Use distraction or progressive muscle relaxation when nauseated.
f. If vomiting occurs, gradually resume oral intake with frequent sips of clear liquids or ice, progressing to bland foods.

Crackers and hard candy often relieve queasiness, whereas hot, spicy, sweet, or strong smelling foods may increase nausea. Alternative nausea relief measures may be effective.

- Provide small feedings of high-kilocalorie, high-protein foods and fluids. *This increases nutritional intake.*
- Assist with oral care, general hygiene, and environmental control of temperature, appearance, and odors. *These measures enhance appetite.*
- Identify and provide preferred foods. *This promotes nutritional intake.*
- Assist to a sitting position during and immediately after meals. *The sitting position helps decrease early feelings of fullness.*

Disturbed Body Image

The diagnosis of cancer is often devastating to the sense of trust in and the perception of one's body. Radiation and chemotherapy lead to changes in appearance and body function (e.g., hair loss, reduced libido, and infertility), further altering body image. Reactions to this diagnosis vary and may include refusal to look in a mirror or discuss the effects of the disease or treatment, unwillingness to participate in rehabilitation, inappropriate treatment decisions, increasing dependence on others or refusal to provide self-care, hostility, withdrawal, and signs of grieving.

- Assess perception of body image through subjective information such as:
  a. What the client likes most and least about his or her body
  b. Preillness perception of people who are sick or have a disability
  c. Current understanding of health and limitations imposed by illness or treatment
  d. Feelings about the illness and its effect on perception of self and others.

  Body image is one's mental idea or picture of the body. It is based on past and present experiences and includes components of one's actual body and emotional responses to that body. Body image changes constantly. There is often a time lag between an actual body change and the changed body image; during this time, the diagnosis, teaching, and treatment may be rejected.
- Discuss the risk for and measures to cope with alopecia. Suggest wearing wigs, scarves, hats, or caps. Teach proper scalp care using baby shampoo or mild soap, a soft brush, sunscreen, and mineral oil to reduce itching. If eyelashes and eyebrows are lost, teach eye protection, such as wearing eyeglasses and caps with wide brims. *Chemotherapeutic agents attack rapidly dividing cells such as those responsible for hair growth. Hair loss usually begins 1 to 2 weeks after initiation of chemotherapy, with maximum loss 1 to 2 months later. Alopecia may range from thinning to total hair loss. Regrowth depends on the treatment schedule and doses; however, it usually begins 2 to 3 months after treatment ends. New hair may be softer, more curly, and slightly different in color. Teaching and emotional support help the client anticipate hair loss, discuss its potential effect on body image, and learn self-care techniques.*
- Discuss available resources for financial assistance with purchase of wigs, including local American Cancer Society chapters and insurance plans. *A well-matched wig (or one the color the client has always wished for!) can help maintain a positive body image.*

Sexual Dysfunction

Sexual dysfunction may result from the malignancy and the effects of radiation and chemotherapy. Reproductive tissues are made of rapidly dividing cells, and cancer treatment may cause temporary or permanent sterility, changes in menstruation, and changes in libido.

- Encourage discussion of actual or potential sexual dysfunction or sterility with the client and significant other. *Clients may be reluctant to discuss this unintended effect of treatment unless encouraged.*
- Assess knowledge, provide information, and clarify misconceptions. Discuss realistic measures for coping (e.g., sperm banking prior to chemotherapy or radiation therapy). *Clients and their partners may be unclear about expected effects on sexuality, reproduction, and the permanency of these effects.*
- Refer for counseling as indicated. *Sexual counseling can help the client and partner develop alternative strategies for expressing their sexuality.*

Risk for Impaired Skin Integrity

Malignant lymphomas may cause significant pruritus and drenching night sweats. As a result, skin integrity may be impaired. In addition, radiation therapy can cause superficial burns, which also may affect skin integrity.

- Frequently assess skin, especially in areas undergoing radiation. *Early identification of lesions allows timely treatment and can prevent further disruption of this important line of defense against infection.*
- Provide and teach measures to promote comfort and relieve itching: Use cool water and a mild soap to bathe; blot (rather than rub) dry skin; apply plain cornstarch or nonperfumed lotion or powder to the skin unless contraindicated; use lightweight blankets and clothing; maintain adequate humidity and a cool room temperature; wash bedding and clothes in mild detergent, and put them through second rinse cycle. *Pruritus is aggravated by excessive warmth, excessive dryness, rough fabrics, fatigue, and stress. Lotions and some powders may be contraindicated during radiation therapy.*

Linking NANDA, NIC, and NOC

Chart 34–3 shows linkages between nursing diagnoses, nursing interventions, and nursing outcomes for the client with malignant lymphoma.

Community-Based Care

When teaching the client and family for home care, include the following topics in addition to those previously identified for specific nursing diagnoses:

- Information about the illness, planned treatment, and anticipated side effects of treatment

NANDA, NIC, AND NOC LINKAGES

CHART 34–3 The Client with Malignant Lymphoma

NANDA

Sexual Dysfunction

↓

NIC

Active Listening

Coping Enhancement

Emotional Support

Sexual Counseling

↓

NOC

Body Image

Psychosocial Adjustment: Life Change

Self-Esteem

Sexual Functioning

Data from *NANDA's Nursing Diagnoses: Definitions & Classification 2005–2006* by NANDA International (2005), Philadelphia; *Nursing Interventions Classification (NIC)* (4th ed.) by J. M. Dochterman & G. M. Bulechek (2004), St. Louis, MO: Mosby; and *Nursing Outcomes Classification (NOC)* (3rd ed.) by S. Moorhead, M. Johnson, and M. Mass (2004), St. Louis, MO: Mosby.

- Skin care and measures to relieve itching and protect areas of radiation
- Symptoms to report to the physician, including those of vertebral compression (decreased sensation or strength in lower extremities)
- Use of analgesics and alternative relief strategies for abdominal pain and peripheral neuropathies
- Respiratory care if mediastinal nodes are enlarged or lungs or pleurae are involved
- Planning activities of daily living to ensure adequate rest and exercise
- Measures to relieve nausea and maintain adequate nutrition.

Refer clients and family members to the local chapter of the American Cancer Society for information, assistance, and counseling. A list of state and local agencies that offer information about malignant lymphoma and financial assistance can be obtained from the Leukemia Society of America.

THE CLIENT WITH MULTIPLE MYELOMA

Multiple myeloma is a malignancy in which plasma cells multiply uncontrollably and infiltrate the bone marrow, lymph nodes, spleen, and other tissues. *Plasma cells* are B-cell lymphocytes that develop to produce antibodies (immunoglobins).

Incidence and Risk Factors

The incidence of multiple myeloma is increasing slightly, with an estimated 15,980 cases diagnosed and 11,300 deaths due to the disease in 2005 (ACS, 2005). It affects blacks nearly twice as often as whites, and men slightly more frequently than women. The incidence of multiple myeloma increases with age, rarely occurring before age 40 (Kasper et al., 2005). Its cause is unknown. Possible contributing factors include genetic alterations, radiation exposure, oncogenic virus, inflammatory stimuli, and chronic antigenic stimulation. The risk for developing multiple myeloma is higher in people of lower socioeconomic status. This increased risk may relate to environmental factors such as poor housing, occupational hazards, poor nutritional status, and other physical and psychosocial stressors such as exposure to infectious agents (Mangan, 2005).

Pathophysiology

Malignant plasma cells arise from one clone (*monoclonal*) of B cells that produce abnormally large amounts of a particular immunoglobin called the *M protein.* This abnormal protein interferes with normal antibody production and impairs the humoral immune response. It also increases blood viscosity and may damage kidney tubules. As myeloma cells proliferate, they replace the bone marrow and infiltrate the bone itself. Cortical bone is progressively destroyed by tumor growth and enzymes produced by myeloma cells. These enzymes facilitate bone destruction, its infiltration by tumor cells, development of new blood vessels to sustain the tumor, and growth of myeloma cells (McCance & Huether, 2006). Affected bones (primarily the vertebrae, ribs, skull, pelvis, femur, clavicle, and scapula) are weakened and may break without trauma (*pathologic fracture*). With disease progression, malignant cells spread via the bloodstream to invade other organs (Figure 34–11 ■).

Manifestations

The disease develops slowly, with up to 30% of clients diagnosed during evaluation for unrelated problems (Mangan, 2005). Manifestations of multiple myeloma are due to its effects on the bone and the impaired immune response due to M-protein production. Bone pain is the most common presenting symptom. With progression of the disease, the pain may increase in severity and become more localized. Rapid bone destruction releases calcium from the bone, leading to hypercalcemia and manifestations of neurologic dysfunction, such as lethargy, confusion, and weakness.

As functional antibody formation decreases and the humoral immune response is suppressed, recurrent infections develop. Cell-mediated immunity remains intact. *Bence Jones proteins* are found in the urine in multiple myeloma. These proteins are toxic to the renal tubules, and may lead to renal failure with azotemia and uremia. (See Chapter 29 ∞ for more information about renal failure.)

About 15% of clients with multiple myeloma die within 3 months of the diagnosis. More frequently, the disease course is chronic, progressing more rapidly with each relapse after remission. The acute terminal stage of the disease is marked by pancytopenia and widespread organ infiltration by myeloma cells (Kasper et al., 2005).

Figure 34–11 ■ An illustration of the progress of multiple myeloma in an African American male. Abnormal plasma cells proliferate uncontrollably, gradually replacing bone marrow and infiltrating bone itself. As the disease progresses, these cells spread to other organs via the bloodstream.

Source: Kevin A. Somerville, Phototake NYC

INTERDISCIPLINARY CARE

Diagnosis and Staging

Diagnostic tests for multiple myeloma include the following:

- *X-rays* and other radiologic studies of the bone may reveal multiple punched-out lesions.
- *Bone marrow examination* shows an abnormal number of immature plasma cells.
- *CBC* shows moderate to severe anemia, and the *ESR* usually is elevated.
- *Protein electrophoresis* shows a spike of one type of antibody, usually IgG.
- *Serum calcium, creatinine, uric acid,* and *BUN* levels often are elevated.
- *Urinalysis* shows Bence Jones protein in the urine.
- *Biopsy* of myeloma lesions confirms the diagnosis of multiple myeloma.

Staging of multiple myeloma is based on the hemoglobin and serum calcium levels, the amount of abnormal protein present, and the degree of bone involvement.

Treatment

There is no cure for multiple myeloma. In some clients, active observation is indicated, as the disease may continue with a slow, *indolent* (sluggish, not developing or progressing) course for many years (Mangan, 2005). When indicated by disease stage or progression, standard treatment includes induction chemotherapy followed by stem cell transplant and maintenance chemotherapy to control progression of the disease. Supportive care is provided to reduce complications of the disease and their effects.

Combination chemotherapy with an alkylating agent (melphalan [Alkeran], cyclophosphamide [Cytoxan], or chlorambucil [Chloromycetin]) and prednisone administered for 4 to 7 days every 4 to 6 weeks is commonly used. Chemotherapy typically reduces bone pain, hypercalcemia, anemia, and the number of infections (Kasper et al., 2005). Localized radiation therapy may be used to treat painful bone lesions. High-dose chemotherapy followed by peripheral allogeneic stem cell transplant may be more effective in achieving a cure, but is associated with a high mortality rate. When autologous SCT is used, granulocyte colony-stimulating factor is administered prior to harvesting and preserving peripheral stem cells for transplant.

Supportive care may include treatment of hypercalcemia with hydration, possible bisphosphonate therapy to reduce bone loss (see Chapter 42 ∞), and calcium, vitamin D, and fluoride supplements to support bone structure. Plasma exchange therapy (plasmapheresis) to remove circulating M proteins is used as needed to treat acute renal failure. Infections are treated promptly when they develop.

NURSING CARE

Assessment

Focused assessment data for the client with multiple myeloma include the following:

- *Health history:* Complaints of back or bone pain, onset, duration, and intensity; complaints of weakness, fatigue, anorexia; history of frequent or recurrent infections; neurologic symptoms such as numbness and tingling or clumsiness.
- *Physical examination:* Level of consciousness and mental status; mobility, gait; localized tenderness or pain, bony crepitus with movement or palpation; movement and sensation in extremities.

Nursing Diagnoses and Interventions

Nursing care of the client with multiple myeloma focuses on problems of chronic pain, impaired mobility, and the risk for injury. Risk for infection is a major nursing care focus; see the previous section on leukemia for specific interventions to reduce this risk. Other nursing care needs are similar to those of clients with other cancers and chronic pain. See Chapters 9 and 14 ∞ for additional specific nursing interventions for these problems.

Chronic Pain

Clients with multiple myeloma typically experience chronic back pain and deep bone pain as myeloma cells saturate the bone marrow and invade the bone structure. Pathologic fractures are a common and reoccurring problem.

- Assess pain, including intensity (use a standard pain scale), onset, duration, precipitating factors, and effective relief

measures. *Identifying the intensity, causes, and precipitating factors of pain helps determine and evaluate effective pain relief measures.*

- Determine position of greatest comfort, and assist as needed into this position. *The client is best able to identify positions that minimize pain, but may need assistance with repositioning.*
- Support position with pillows. *Bony prominences may be painful due to infiltrates. Pillows can help relieve pressure on these prominences, thus reducing pain.*
- Provide uninterrupted rest periods. *Adequate rest facilitates pain relief and improves pain tolerance.*
- Teach adjunctive pain relief strategies such as relaxation or guided imagery. *A combination of pharmacologic and nonpharmacologic methods provides better management of chronic pain, especially bone pain.*
- Teach effective analgesic use, including the family in instruction. *Analgesics are most effective when taken before pain becomes severe. Clients and their families may be reluctant to use prescription analgesics on a regular basis.*
- Report unrelieved pain to the physician. *A different analgesic or addition of an adjunctive medication such as a nonsteroidal anti-inflammatory drug (NSAID) may be needed to effectively control pain.*

Impaired Physical Mobility

Painful bony infiltrates and pathologic fractures may limit mobility. A brace or splint may be used to protect extremities or support the back. In addition, persistent weakness associated with the cancer and anemia may limit the client's ability to participate in usual activities.

- Assist to change position at least every 2 hours. *Assistance with repositioning is necessary due to weakness. Frequent repositioning improves comfort and reduces the risk for impaired skin and tissue integrity.*

PRACTICE ALERT

Gently support extremities during repositioning. Weakened extremities due to infiltration of bone by myeloma cells and muscle atrophy from lack of use increase the risk for pathologic fractures.

- Provide a trapeze to assist in repositioning. *A trapeze provides better leverage, allowing the client to assist with repositioning and providing a degree of independence. The ability to participate in self-care improves self-esteem.*

Risk for Injury

The bone involvement of multiple myeloma places the client at high risk for pathologic and traumatic fractures. Pathologic fractures can occur with simple activities such as turning or reaching for an item. The spine usually is affected; the ribs and bones of the extremities also may be at risk for fracture.

- Place needed items close at hand. *Straining to reach objects increases the risk of falling or sustaining other injury.*
- Provide safety measures to prevent falls from bed: Place the bed in a low position, use side rails as indicated, and place the call bell within reach. *Safety measures help prevent accidental injury. A secure environment minimizes risk and helps prevent falls.*
- Provide shoes with nonskid soles, a clear pathway, adequate lighting, and a level surface free of scatter rugs or other hazards when ambulating. Provide a walker as needed for support and security. *Weight-bearing exercise promotes bone repair. Safety measures, such as an unobstructed pathway and a firm walking surface, help prevent falls.*

Community-Based Care

When teaching clients and their families for home care, include the following topics:

- Strategies for home maintenance management
- Signs and symptoms of complications to be reported to the physician (e.g., symptoms of vertebral and extremity fractures)
- Manifestations of infection to report: fever and chills; increased malaise, fatigue, or weakness; cough with or without sputum; sore throat; dysuria, nocturia, frequency, urgency, or malodorous urine.

Provide referrals for home health and home maintenance services, physical or occupational therapy, social services, and hospice care as appropriate.

THE CLIENT WITH NEUTROPENIA

Leukopenia is a decrease in the total circulating white blood cell count. Although any type of WBC may be affected, neutrophils, which make up the majority of WBCs, are affected most often. *Neutropenia* is a decrease in circulating neutrophils, usually less than 1500 cells/mm^3. Neutropenia may be either congenital or acquired, developing secondarily to prolonged infection, hematologic disorders, starvation, or autoimmune disorders (such as rheumatoid arthritis). Chemotherapy and other drugs can suppress the bone marrow. Neutropenia develops in approximately half of clients undergoing chemotherapy to treat cancer (Eggenberger et al., 2004). *Agranulocytosis* is severe neutropenia, with less than 200 cells/µm. Numbers of other granulocytes also are reduced. It is usually due to impaired leukocyte formation in the bone marrow or increased cell destruction in circulating blood. Agranulocytosis significantly increases the risk for infection. *Aplastic anemia* affects production of all blood cells, resulting in anemia, thrombocytopenia, and agranulocytosis.

Pathophysiology and Manifestations

Neutrophils are an integral component of the immune response. They are phagocytes, drawn to and activated by infection and inflammation to engulf and degrade invading microorganisms. Their life span in peripheral blood is short, less than 1 day. When *granulopoiesis* (the development and maturation of granulocytes) in the bone marrow is suppressed, the number of circulating neutrophils falls rapidly. As a result, the body's ability to defend itself against infection is significantly reduced.

The manifestations of neutropenia reflect the resulting impaired immunity and inflammatory response. Opportunistic

bacterial, fungal, and protozoal infections develop, commonly affecting the respiratory tract and mucosa of the mouth, GI tract, and vagina. Malaise, chills, and fever with extreme weakness and fatigue are common manifestations.

INTERDISCIPLINARY CARE

The diagnosis of neutropenia is made based on the client's manifestations, risk factors, and the CBC. The total white blood count is low, often less than 1000/mm^3.

Hematopoietic growth factors such as GM-CSF are administered to stimulate granulocyte maturation and differentiation. Infections are treated with antibiotic therapy. Protective isolation procedures may be initiated to prevent exposure to pathogens. When neutropenia is related to chemotherapy, cancer treatment often must be halted, at least temporarily, to allow the bone marrow to recover.

NURSING CARE

The primary nursing care focus is early identification of neutropenia and protecting the client from infection. The WBC count is monitored on a regular basis, and any decline reported to the physician. Protective isolation may be indicated, including restricting the number of visitors and people with apparent illness. See *Risk for Infection* in the earlier section on leukemia for specific nursing interventions for the client with neutropenia.

THE CLIENT WITH INFECTIOUS MONONUCLEOSIS

Infectious mononucleosis is characterized by invasion of B cells in the oropharyngeal lymphoid tissues by the Epstein-Barr virus. This disease is usually benign and self-limiting. It often affects young adults between the ages of 15 and 30. Although many children are infected with EBV, symptomatic infectious mononucleosis is uncommon in early childhood. The virus is present in saliva, which appears to be the primary mode of transmission. As a result, infectious mononucleosis is often called the "kissing disease."

EBV also is associated with some cancers, including Burkitt's lymphoma and Hodgkin's disease, B-cell lymphoma, and nasopharyngeal carcinoma (Kasper et al., 2005).

Pathophysiology and Manifestations

When the virus enters the body, unaffected B cells produce antibodies against the virus, and T cells directly attack the virus. Infected B cells are destroyed as the virus replicates. The proliferation of B and T cells, as well as the removal of dead and damaged leukocytes, is responsible for the swelling of lymphoid tissues.

The incubation period for infectious mononucleosis is 4 to 8 weeks. Its onset is insidious, with headache, malaise, and fatigue. Fever, sore throat, and cervical lymphadenopathy (lymph node enlargement and pain) lasting 1 to 3 weeks is common. Symptom severity varies from person to person. Lymph node involvement may be generalized; about 50% of people with infectious mononucleosis develop an enlarged spleen (splenomegaly).

INTERDISCIPLINARY CARE

Laboratory findings include increased lymphocytes and monocytes, with about 20% of the cells atypical in form. Early in the infection, the WBC count usually is normal or low, but by the second week it increases and remains elevated for 4 to 8 weeks. Platelet counts are often low during the illness.

Recovery occurs in 2 to 3 weeks; however, debility and lethargy may last for up to 3 months. The treatment includes bed rest and analgesic agents to alleviate the symptoms. Nursing care is primarily educational to prevent further spread of the disease.

PLATELET AND COAGULATION DISORDERS

Platelet and coagulation disorders affect **hemostasis**, control of bleeding. Hemostasis maintains a relatively steady state of blood volume, blood pressure, and blood flow through injured vessels. Bleeding disorders result from deficient platelets, disruption of the clotting cascade, or a combination of factors

THE CLIENT WITH THROMBOCYTOPENIA

Thrombocytopenia is a platelet count of less than 100,000 per milliliter of blood. It can lead to abnormal bleeding. A continuing decline in circulating platelets to less than 20,000/mL can lead to spontaneous bleeding and hemorrhage from minor trauma (Figure 34–12 ■). Bleeding due to platelet deficiency usually occurs in small vessels, causing manifestations such as *petechiae* and *purpura*. The mucous membranes of the nose, mouth, GI

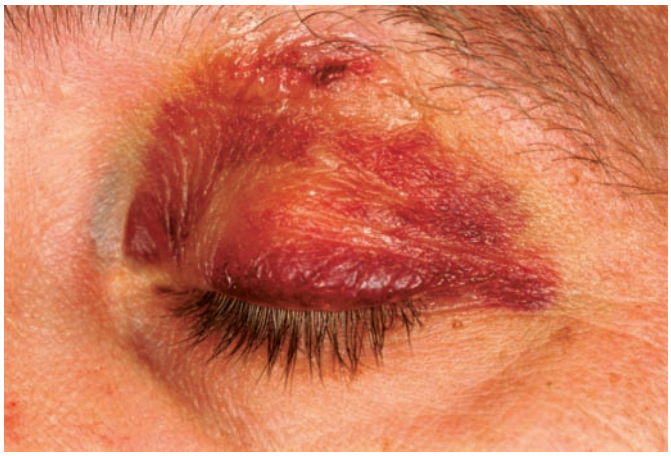


Figure 34–12 ■ Significant ecchymosis of the eyelid associated with minor trauma in a client with thrombocytopenia.

Source: Scott Camazine, Photo Researchers, Inc.

tract, and vagina often bleed. Serious and potentially fatal bleeding occurs when the platelet count is less than 10,000/mL.

Thrombocytopenia results from one of three mechanisms: decreased production, increased sequestration in the spleen, or accelerated destruction. Primary thrombocytopenia that leads to increased platelet destruction is discussed below. Secondary thrombocytopenia may be caused by aplastic anemia, bone marrow malignancy, infection, radiation therapy, or drug therapy (Box 34–6). Heparin therapy is the most common drug-induced thrombocytopenia; it is included in the discussion that follows. Platelet sequestration usually is due to an enlarged spleen. Up to 80% of platelets may be removed from circulation with significant splenomegaly (Porth, 2005). Finally, thrombocytopenia may result from premature platelet destruction associated with disseminated intravascular coagulation (DIC).

Physiology Review

Effective control of bleeding requires a series of complex interactions between the damaged tissue and blood vessel, platelets, clotting factors, and processes to dissolve clots once bleeding has been controlled (see Figures 33–5, 33–6, and 33–7 ∞). Platelets are formed in the bone marrow under control of thrombopoietin, a protein produced by the liver, kidney, smooth muscle, and bone marrow. Platelets are attracted to the damaged vessel wall, where they aggregate and release mediators that activate the clotting process. See Chapter 33 ∞ for a more complete discussion about platelets, clotting, and hemostasis.

Pathophysiology

The two types of primary thrombocytopenia are immune thrombocytopenic purpura and thrombotic thrombocytopenic purpura.

Immune Thrombocytopenic Purpura

Immune thrombocytopenic purpura (ITP), also known as *idiopathic thrombocytopenic purpura,* is an autoimmune disorder in which platelet destruction is accelerated. In ITP, proteins on the platelet cell membrane stimulate autoantibody production, usually IgG antibodies. These autoantibodies adhere to the platelet membrane. Although the platelets function normally, the spleen reacts to them as being foreign and destroys the altered platelets after only 1 to 3 days of circulation.

MANIFESTATIONS The manifestations of ITP are due to bleeding from small vessels and mucous membranes. Petechiae and purpura develop, often on the anterior chest, arms, neck, and oral mucous membranes. Bruising also may be apparent. As bleeding progresses, epistaxis (nosebleed), hematuria, excess menstrual bleeding, and bleeding gums occur. Spontaneous intracranial bleeding is rare but does occur. Associated symptoms include weight loss, fever, and headache.

INCIDENCE AND COURSE Acute ITP affects people of any age following a viral illness (Copstead & Banasik, 2005). Acute ITP typically lasts only 1 to 2 months, resolving without long-term consequences. In its chronic form, ITP typically affects adults between ages 20 and 50; women are affected more often than men. Its onset is insidious. Chronic (or adult) ITP often occurs in people with other immune-associated disorders such as systemic lupus erythematosus or HIV disease (Copstead & Banasik, 2005).

Thrombotic Thrombocytopenic Purpura

Thrombotic thrombocytopenic purpura (TTP) is a rare disorder in which thrombi occlude arterioles and capillaries of the microcirculation. Many organs are affected, including the heart, kidneys, and brain. The incidence of TTP is increasing (McCance & Huether, 2006). Its cause is unknown. Platelet aggregation is a key feature of the disorder. As RBCs circulate through partially occluded vessels, they fragment, leading to hemolytic anemia (Porth, 2005).

MANIFESTATIONS TTP may be acute, the more common and severe form, or chronic. Acute idiopathic TTP may be fatal within months if untreated. The manifestations of TTP include purpura and petechiae, and neurologic symptoms such as headache, seizures, and altered consciousness.

Heparin-Induced Thrombocytopenia

Heparin-induced thrombocytopenia (HIT) develops as a result of an abnormal response to heparin therapy. Unfractionated heparin carries a greater potential to precipitate HIT; it can, however, develop in clients receiving low-molecular-weight heparin who have previously been treated with unfractionated heparin. See Chapter 35 ∞ for further discussion of heparin therapy and the forms of heparin.

Heparin is a protein that occurs naturally in human tissues and inflammatory cells. It can react directly with platelets, causing them to agglutinate (clump) and be removed from circulation by phagocytosis. This form of HIT, called type I HIT, typically causes mild thrombocytopenia. The more severe form, type II HIT, results from an immune reaction to heparin. In type II HIT, heparin forms an immune complex with a platelet protein known as platelet factor 4 (PF4). This complex acts as a foreign antigen in some clients, stimulating antibody production. The antibody binds with the heparin-PF4 complex, and these antibody-heparin-PF4 complexes subsequently bind with circulating

BOX 34–6 Selected Causes of Secondary Thrombocytopenia

Diseases
- Vitamin B_{12} anemia
- Folic acid anemia
- Aplastic anemia
- Leukemia
- Alcoholism
- DIC
- Infectious mononucleosis
- Viral infections
- HIV disease

Drugs
- Thiazide diuretics
- Aspirin
- Ibuprofen
- Indomethacin
- Naproxen
- Sulfonamides
- Phenytoin
- Cimetidine
- Digoxin
- Furosemide
- Heparin
- Morphine

Treatments
- Radiation therapy
- Chemotherapy
- Massive transfusion of stored blood

MEDIALINK Case Study: Immune Thrombocytopenic Purpura

platelets, causing them to aggregate. As affected platelets aggregate, they are removed from circulation, leading to thrombocytopenia. In addition, small pieces of platelets can break loose, stimulating the clotting cascade and the development of thrombosis (clotting) (Francis & Drexler, 2005). The thrombocytopenia and the thrombosis can be reversed by prompt withdrawal of heparin therapy (Kasper et al., 2005).

MANIFESTATIONS Despite thrombocytopenia, bleeding is usually a manifestation of HIT, probably because of the increased tendency to form clots that deplete clotting factors. The client may develop manifestations of an arterial thrombosis (severe pain, paresthesias, pallor and cool skin temperature, and pulselessness distal to the arterial occlusion) or of venous thrombosis (edema, redness, and warmth of the affected area). On rare occasion, an intravenous bolus of unfractionated heparin can precipitate an acute inflammatory response with manifestations that may mimic an acute pulmonary embolism: fever, chills, hypertension, tachycardia, dyspnea, chest pain, and cardiopulmonary arrest (Francis & Drexler, 2005).

INTERDISCIPLINARY CARE

The diagnosis of thrombocytopenia is based on history, manifestations, and diagnostic test results. Management focuses on treating or removing any causative factors and treating the platelet deficiency.

Diagnosis

The following diagnostic tests are used to identify thrombocytopenia:

- *CBC with platelet count* is done to evaluate blood cell counts, hemoglobin, and hematocrit.
- *Antinuclear antibodies (ANA)* are measured to assess for autoantibodies and identify possible contributing disorders such as systemic lupus erythematosus.
- *Serologic studies* for hepatitis viruses, cytomegalovirus (CMV), EBV, toxoplasma, and HIV may be done. Serologic testing also may be performed when HIT is suspected.
- *Bone marrow examination* evaluates for aplastic anemia and megakaryocyte production.

Medications

Oral glucocorticoids, such as prednisone, are prescribed to suppress the autoimmune response. Many clients who respond to glucocorticoid treatment relapse when the drug is withdrawn, however. Immunosuppressive drugs such as azathioprine, cyclophosphamide, and cyclosporine may be used.

Prompt withdrawal of heparin therapy is vital when HIT is the cause of thrombocytopenia. All sources of heparin are removed, including heparin used to flush intravenous or other catheters and heparin-coated catheters (Francis & Drexler, 2005). A nonheparin anticoagulant such as lepirudin (Refludan) or argatroban may be substituted. Lepirudin is a thrombin inhibitor. It is a recombinant form of hirudin, originally isolated from the salivary glands of leeches. Its primary adverse effect is bleeding; as a protein, it also can stimulate antibody development, resulting in rare instances of anaphylaxis. Argatroban is a synthetic direct thrombin inhibitor with a short half-life. It clears quickly when the infusion is discontinued, an advantage if excessive bleeding develops or invasive procedures must be performed (Rice et al., 2002).

Treatments

Platelet transfusions may be required to treat acute bleeding due to thrombocytopenia. Platelets are prepared from fresh whole blood; one unit contains 30 to 60 mL of platelet concentrate. The expected increase in platelets after one unit is infused is 10,000/mL. *Plasmapheresis,* or *plasma exchange therapy,* is the primary treatment for acute thrombotic thrombocytopenic purpura. The client's plasma is removed and replaced with fresh frozen plasma to remove autoantibodies, immune complexes, and toxins.

Surgery

A *splenectomy* (surgical removal of the spleen) is the treatment of choice if the client with ITP relapses when glucocorticoids are discontinued. The spleen is the site of platelet destruction and antibody production. This surgery often cures the disorder, although relapse may occur years after splenectomy.

NURSING CARE

Assessment

- *Health history:* Complaints of bruising with minor or no trauma, bleeding gums, nosebleed, heavy or prolonged menstrual periods, black, tarry, or bloody stools, hematemesis, headache, fever, or neurologic symptoms; recent weight loss; recent viral or other illness; current and recent medications; exposure to toxins; previous exposure to heparin.
- *Physical examination:* Skin and mucous membranes for color, temperature, petechiae, purpura, or bruises; vital signs; weight; mental status and level of consciousness; heart and breath sounds; abdominal exam; body fluids for occult blood.
- *Diagnostic tests:* CBC, hemoglobin and hematocrit, platelet count; serologic and ANA test results; bone marrow examination results.

Nursing Diagnoses and Interventions

Inadequate platelets impair hemostasis, placing the client at risk for bleeding. Bleeding gums, an early sign of the disorder, affects oral mucous membrane integrity as well.

Ineffective Protection

Bleeding is a serious complication associated with thrombocytopenia. As platelet counts (measured in cubic millimeters) decrease, the risk of bleeding increases. The risk is minimal with counts greater than 50,000 mm^3; moderate when the count is between 20,000 and 50,000 mm^3; and significant when the count falls below 20,000 mm^3.

- Monitor vital signs, heart and breath sounds every 4 hours. Frequently assess for other manifestations of bleeding:
  a. Skin and mucous membranes for petechiae, ecchymoses, and hematoma formation
  b. Gums, nasal membranes, and conjunctiva for bleeding
  c. Overt or occult blood in emesis, urine, or stool

d. Vaginal bleeding
e. Prolonged bleeding from puncture sites
f. Neurologic changes: headache, visual changes, altered mental status, decreasing level of consciousness, seizures
g. Abdominal: epigastric pain, absence of bowel sounds, increasing abdominal girth, abdominal guarding or rigidity.

Early identification of bleeding is important to prevent serious blood loss and shock.

PRACTICE ALERT

Avoid invasive procedures such as rectal temperatures, urinary catheterization, and parenteral injections to the extent possible. Diagnostic procedures such as biopsy or lumbar puncture should be avoided if the platelet count is less than 50,000 mm^3. Invasive procedures can cause tissue trauma and bleeding. Procedures that use large-bore needles should be delayed until the platelet count is increased.

- Apply pressure to puncture sites for 3 to 5 minutes; apply pressure to arterial puncture sites for 15 to 20 minutes. *Pressure promotes hemostasis and clot formation.*
- Instruct to avoid forcefully blowing the nose or picking crusts from the nose, straining to have a bowel movement, and forceful coughing or sneezing. *These activities increase the risk of external and internal bleeding.*

Impaired Oral Mucous Membranes

Thrombocytopenia frequently leads to bleeding of the gums and oral mucosa. As a result, risk for infection and impaired nutrition increases.

- Frequently assess the mouth for bleeding. Inquire about oral pain or tenderness. *Breakdown of oral mucous membranes increases the risk of infection and bleeding, and causes discomfort with eating.*
- Encourage use of a soft-bristle toothbrush or sponge to clean teeth and gums. *Hard bristles may abrade oral mucosa, causing bleeding and increasing the risk of infection.*
- Instruct to rinse the mouth with saline every 2 to 4 hours. Apply petroleum jelly to lips as needed to prevent dryness and cracking. *Saline mouth rinses and petroleum jelly help maintain oral tissue integrity and promote cleansing and healing.*
- Instruct to avoid alcohol-based mouthwashes, very hot foods, alcohol, and crusty foods. Teach to drink cool liquids at least every 2 hours. *Avoiding foods and liquids that traumatize oral mucosa increases comfort; fluid intake prevents dehydration and helps maintain mucous membrane integrity.*

Community-Based Care

In the adult, ITP often is a chronic disorder that the client and family must learn to manage. Secondary thrombocytopenia may be either acute or chronic. Discuss the following topics when preparing the client and family for home care:

- Nature of the disorder, its usual course, and the treatment plan
- Use of and desired and potential adverse effects of prescribed medications
- Risks and benefits of surgery or treatments such as plasma replacement therapy
- The importance of follow-up tests and visits for care
- Measures to reduce the risk of bleeding: safety measures such as a soft-bristle toothbrush, electric razor, avoidance of contact sports and hazardous activities, and avoiding medications that further interfere with platelet function (Box 34–7)

Refer for home health or other community services (e.g., housekeeping, shopping) as indicated.

THE CLIENT WITH HEMOPHILIA

Hemophilia is a group of hereditary clotting factor disorders that lead to persistent and sometimes severe bleeding (see the accompanying Genetic Considerations box). Although often considered a disease of children, hemophilia may be diagnosed in adults. Deficiencies of three clotting factors, VIII, IX, and XI, account for 90% to 95% of the bleeding disorders collectively called hemophilia (McCance & Huether, 2006).

Physiology Review

When tissue injury occurs, platelets collect at the site, adhering to the damaged vessel wall (the platelet plug). Activation of the clotting cascade, a sequential process of interactive reactions of clotting factors, is vital to form a stable clot. Clotting factors are plasma proteins primarily produced by the liver. A number of these factors require the presence of vitamin K for synthesis and activation. Once the clot has been formed and stabilized, it begins to retract, pulling together the edges of the damaged blood vessel to initiate the healing process.

Pathophysiology

Hemophilia A (or *classic hemophilia*) is the most common type of hemophilia, caused by deficiency or dysfunction of clotting factor VIII. It is transmitted as an X-linked recessive disorder from mothers to sons (Figure 34–13 ■). The genetic defect of hemophilia A on the X chromosome may cause deficient factor VIII production or a defective form of the pro-

BOX 34–7 Medications That May Interfere with Platelet Function

Over-the-Counter Medications

- Aspirin and salicylates, including:
  - Alka-Seltzer
  - Bufferin
  - Doan's pills
  - Ecotrin
  - Excedrin
  - Midol
  - Pepto-Bismol
  - Vanquish
- NSAIDs such as
  - Advil
  - Aleve
  - Nuprin
  - Pamprin IB

Prescription Medications

- Aspirin-containing analgesics
- Chemotherapy drugs
- Antibiotics such as penicillin
- Carbamazepine (Tegretol)
- Colchicine
- Dipyridamole (Persantine)
- Gold salts
- Heparin
- Quinine derivatives
- Sulfonamides
- Thiazide diuretics

GENETIC CONSIDERATIONS
Focus on Hemophilia

The incidence and pattern of inheritance for the forms of hemophilia differ.

- Hemophilia A occurs in about 1 in 10,000 male births, transmitted on the X chromosome: Each male offspring has a 50% risk of inheriting the defective gene; each female offspring has a 50% risk of becoming a carrier.
- Hemophilia B occurs in about 1 in 100,000 male births, transmitted on the X chromosome.
- Von Willebrand's disease affects about 1 in 100 to 500 people, usually inherited as an autosomal dominant trait: Offspring of an affected person have a 50% risk of inheriting the trait and the disorder.
- Factor XI deficiency inherited as an autosomal recessive trait: Each offspring of a carrier and an unaffected individual has as 50% risk of inheriting the trait; each offspring of two carriers has a 50% risk of being a carrier and a 25% risk of having the disorder. This deficiency is common in Ashkenazi Jews.

tein. When the concentration of the clotting factor is 5% to 35% of normal, the disease is *mild.* Bleeding is infrequent, and usually associated with trauma. Concentrations of 1% to 5% of normal result in *moderate* disease. Again, bleeding usually occurs secondarily to trauma. *Severe* hemophilia occurs when concentrations are less than 1% of normal. Bleeding is frequent, often occurring without trauma (Kasper et al., 2005; McCance & Huether, 2006).

Hemophilia B (also called *Christmas disease*) accounts for about 15% of cases, and is caused by a deficiency in Factor IX. Despite the difference in clotting factor deficits, hemophilia A and B are clinically identical.

Von Willebrand's disease, often considered a type of hemophilia, is the most common hereditary bleeding disorder (Porth, 2005). It is caused by a deficit of or defective von Willebrand (vW) factor, a protein that mediates platelet adhesion (Tierney et al., 2005). Reduced levels of Factor VIII often also are present, because vW factor carries Factor VIII. This clotting disorder affects men and women equally. Bleeding associated with von Willebrand's disease rarely is severe. It often is diagnosed when prolonged bleeding follows surgery or a dental extraction.

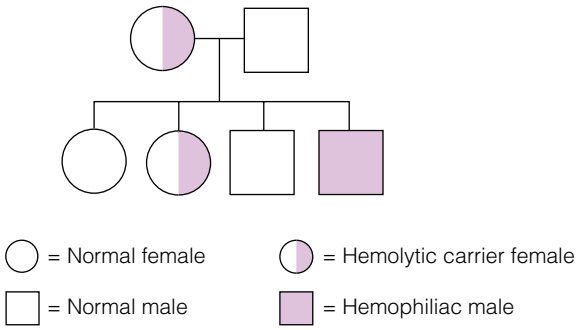


Figure 34–13 ■ The inheritance pattern of hemophilia A and B. Both are transmitted as X-linked recessive disorders. Females may be carriers, but males develop these disorders.

Factor XI deficiency (or *hemophilia C*) is usually a mild disorder, identified when postoperative bleeding is prolonged. A comparison of the types of hemophilia is found in Table 34–9.

People with hemophilia form a platelet plug at the site of bleeding, but the clotting factor deficit impairs formation of a stable fibrin clot. The effect of vW factor deficiency is somewhat different, in that platelet aggregation at the site of injury is impaired. In either case, prolonged or extensive bleeding may result. Often bleeding occurs in response to injury or as a result of surgery. However, a severe clotting factor deficit can lead to spontaneous bleeding into the joints (*hemarthrosis*), deep tissues, and CNS. Hemarthrosis often causes joint deformity and disability, usually of the elbows, hips, knees, and ankles.

Manifestations

The following are manifestations of hemophilia:

- Hemarthrosis
- Easy bruising and cutaneous hematoma formation with minor trauma (e.g., an injection)
- Bleeding from the gums and prolonged bleeding following minor injuries or cuts

TABLE 34–9 **Types of Hemophilia**

TYPE/NAME	DEFICIENCY	CHARACTERISTICS	TREATMENT
Hemophilia A (classic hemophilia)	Factor VIII	Transmitted by females; occurs primarily in males; bleeding time normal; coagulation time prolonged	Factor VIII concentrate or cryoprecipitate
Hemophilia B	Factor IX	Transmitted by females; occurs primarily in males; bleeding time normal; coagulation time prolonged	Factor IX (Christmas disease concentrate)
Von Willebrand's disease	vW factor Factor VIII	Occurs in both females and males; bleeding time and coagulation time are both prolonged	Cryoprecipitate and DDAVP
Factor XI deficiency	Factor XI	Occurs in both males and females; the activated partial thromboplastin time is prolonged	Fresh frozen plasma

- Gastrointestinal bleeding, with hematemesis (vomiting blood), occult blood in the stools, gastric pain, or abdominal pain
- Spontaneous hematuria or epistaxis (nosebleed)
- Pain or paralysis due to the pressure of hematomas on nerves
- Intracranial hemorrhage is a potentially life-threatening manifestation of hemophilia.

INTERDISCIPLINARY CARE

Treatment of hemophilia focuses on preventing and/or treating bleeding, primarily by replacing deficient clotting factors. Specific treatment depends on the severity of the disorder and the specific factor deficiency. Care may be complicated by hepatitis or HIV disease in people with hemophilia treated with clotting factor concentrates prepared from multiple units of donated blood. Today, routine testing of all blood, improved blood donor screening, and current methods of treating hemophilia have significantly reduced the risk for these bloodborne diseases.

Diagnosis

The following laboratory tests may be ordered:

- *Serum platelet levels* are measured and are usually normal.
- *Coagulation studies* such as APTT, bleeding time, and prothrombin time are used to screen for hemophilia when abnormal bleeding occurs. APTT is increased in all types of hemophilia. Prothrombin time is unaffected in these disorders but may be measured to rule out other disorders. Bleeding time is prolonged in von Willebrand's disease but normal in hemophilia A and B.
- *Factor assays* are performed; Factor VIII is decreased in hemophilia A and often in von Willebrand's disease, Factor IX is decreased in hemophilia B, and Factor XI in hemophilia C.
- *Amniocentesis* or *chorionic villus sampling* is used to identify the genetic defect of hemophilia when there is a known family history of the disease.

Medications

Deficient clotting factors are replaced regularly, as a prophylactic measure before surgery and dental procedures, and to control bleeding. Clotting factors may be given as fresh-frozen plasma, cryoprecipitates, or concentrates. Factor levels are measured on a regular basis to determine whether the treatment is adequate. Clotting factors are often self-administered and may be taken either on a regular or intermittent schedule.

Fresh-frozen plasma replaces all clotting factors (including both Factor VIII and Factor IX) except platelets. When the cause of bleeding is not yet determined, fresh-frozen plasma may be administered intravenously until a definitive diagnosis is made.

Hemophilia A is usually treated with either heat-treated Factor VIII concentrate (heat treating reduces the risk of transmitting disease) or recombinant Factor VIII. Although recombinant Factor VIII, produced using recombinant DNA technology, eliminates the risk of viral disease transmission, its use is limited by cost. The dose of Factor VIII is determined by the severity of the deficit and the presence or prospect of active bleeding (e.g., planned surgery).

Desmopressin acetate (DDAVP, Stimate) may be given to people with mild hemophilia A or von Willebrand's disease prior to minor surgeries. This drug causes release of Factor VIII and will raise blood levels by two- or threefold for several hours, reducing the risk of bleeding and the need for clotting factor concentrate (Tierney et al., 2005).

Factor IX concentrate (administered intravenously) is used to treat hemophilia B. Because Factor IX concentrates also contain a number of other proteins, there is risk of thrombosis with recurrent use. They are used judiciously, only when needed. Products produced by recombinant technology or that are monoclonally purified carry a lower risk of stimulating thrombus formation (Kasper et al., 2005). Fresh-frozen plasma replaces Factor XI and is used when necessary. It may be given daily until the risk for bleeding decreases.

Factor VIII concentrates contain functional vW factor, and may be used to treat von Willebrand's disease. Aspirin is avoided in all types of hemophilia.

NURSING CARE

Although primary responsibility for care falls to the client and family, nursing care presents challenges. For additional assessment and nursing care strategies, see the accompanying Nursing Care Plan.

Health Promotion

Encourage clients with a family history of hemophilia or bleeding disorders to seek genetic counseling during their family planning process. Although tests are available for the hemophilia gene, the technology to correct the disorder *in utero* does not yet exist. See Chapter 8 ∞.

Assessment

While severe hemophilia usually is diagnosed in childhood, milder cases may not be identified until surgery, invasive dental work, or a traumatic injury causes extensive or prolonged bleeding. Focused assessment related to hemophilia includes the following:

- *Health history:* Previous bleeding episodes with or without trauma; history of easy bruising, hematomas, epistaxis, bleeding gums, hematuria, vomiting blood, or joint pain; aspirin use; family history of hemophilia or bleeding disorders.
- *Physical examination:* Vital signs; bruising or bleeding of skin or mucous membranes; mental status; abdominal assessment; presence of joint deformity, decreased range of motion.
- *Diagnostic tests:* CBC including hemoglobin, hematocrit, and platelet count; clotting factor assays; tests for occult blood (urine, stool, emesis); x-ray and scan results for evidence of bleeding.

Nursing Diagnoses and Interventions

Impaired blood clotting, the need for continuing care and disease management, and the risk for genetic transmission of hemophilia are priority problems for the client with hemophilia.

NURSING CARE PLAN A Client with Hemophilia

Jermiel Cruise is a 20-year-old student at the community college. He is admitted to the emergency department with a nosebleed that began when he fell during a touch football game. It has continued to bleed for over an hour.

ASSESSMENT

Mr. Cruise states that he has hemophilia and realizes that playing contact sports "is probably a dumb thing to do." He adds that he has not had any recent bleeding episodes. An icebag and manual pressure are applied in the emergency department. The physician orders Factor VIII concentrate to be administered. Physical assessment findings are T 97.2°F (36.2°C), BP 118/64, R 18. Skin pale but warm. Laboratory tests reveal a prolonged APTT and a normal bleeding time and PT. Following treatment, Mr. Cruise's bleeding subsides.

DIAGNOSES

- *Risk for Aspiration* related to uncontrolled nosebleed
- *Noncompliance* with activity recommendations
- *Ineffective Protection* related to lack of clotting factor VIII

EXPECTED OUTCOMES

- Exhibit no further signs of bleeding.
- Maintain vital signs within his usual range.
- Maintain an open airway.
- Identify sports and recreation activities in which he can safely participate.
- Verbalize self-care measures to control bleeding.

PLANNING AND IMPLEMENTATION

- Monitor vital signs and for further signs of bleeding.
- Assess airway and auscultate breath sounds.
- Review emergency measures to help stop bleeding.
- Reiterate the importance of seeking prompt medical attention if bleeding should occur.
- Advise regarding the importance of wearing a Medic-Alert bracelet identifying him as a hemophiliac.
- Discuss alternative noncontact sports and recreational activities.

EVALUATION

On discharge, Mr. Cruise has no further signs of bleeding, shock, or aspiration. He is able to verbalize methods to help stop local bleeding and the importance of seeking medical attention promptly when bleeding continues. Mr. Cruise agrees to stop at a local drug store on the way home to order a Medic-Alert bracelet. In addition, Mr. Cruise verbalizes an understanding of the importance of avoiding contact sports and has identified swimming and golf as alternative leisure activities that he might enjoy.

CRITICAL THINKING IN THE NURSING PROCESS

1. What is the pathophysiologic basis for the bleeding that occurs in hemophilia A and B?
2. What was Mr. Cruise's priority nursing diagnosis? Why?
3. Why is family planning a special consideration with a client who has hemophilia?
4. Outline a plan to teach the family of a client diagnosed with hemophilia how to administer an intravenous infusion.
5. Develop a care plan for Mr. Cruise for the nursing diagnosis *Impaired Social Interaction.* Consider Mr. Cruise's age and developmental level in creating the plan.

See Evaluating Your Response in Appendix C.

Ineffective Protection

The inability to form stable clots and stem bleeding from injured blood vessels creates a significant risk for the client with hemophilia. Nursing care measures focus on preventing injury and protecting the skin from damage.

- Monitor for signs of bleeding, including hematomas, ecchymoses, and purpura, as well as surface oozing or bleeding. Check emesis and stool for occult blood. *Bleeding may occur in cutaneous tissues as well as internal organs. Bleeding in the upper gastrointestinal tract may not be readily apparent in the stool.*
- Notify the physician of any apparent bleeding. *Prompt intervention with administration of clotting factor concentrate decreases the risk of hemorrhage and subsequent hypovolemia.*
- Avoid intramuscular injections, rectal temperatures, and enemas. *These can pose a risk of tissue and vascular trauma, which can precipitate bleeding.*
- Use safety measures in personal care. For example, use an electric razor rather than a razor blade to shave. *Use of an electric razor minimizes the opportunity to develop superficial cuts that may result in bleeding.*
- If bleeding occurs, control blood loss using gentle pressure, ice, or a topical hemostatic agent, such as an absorbable gelatin sponge, microfibrillar collagen hemostat, or topical thrombin. *Direct pressure occludes bleeding vessels. Ice, a vasoconstrictor, may facilitate bleeding control, as do topical hemostatic agents.*
- Instruct to avoid activities that increase the risk of trauma, including contact sports, physical exertion associated with job performance, and to eliminate safety hazards in the home. *Depending on the severity of the clotting factor deficit, even minor trauma can lead to serious bleeding episodes. Safer activities such as noncontact sports (e.g., swimming, golf) and occupations that do not require physical labor may be substituted.*

Risk for Ineffective Health Maintenance

Hemophilia is a chronic disorder, requiring active management to prevent and control bleeding and complications. Frequent visits to the physician or clinic may be necessary. In addition, the client may need to learn to self-administer clotting factors and measures to prevent complications. The lifelong nature of the disorder may interfere with compliance, especially during early adulthood.

- Assess knowledge of disorder and the related treatments. *Assessment allows identification of knowledge gaps and provides a basis on which to provide additional information. Impaired disease management may be due to lack of knowledge or a conscious decision not to follow the recommendations of the healthcare provider.*

- Provide information about the bleeding disorder and prescribed medications and treatments. *Individualized instruction is more effective than general, possibly irrelevant information.*
- Provide emotional support, expressing confidence in the client's self-care abilities. *Emotional support helps the client incorporate the care regimen into his or her lifestyle.*
- Provide supervised learning and practice opportunities for administering clotting factors and topical hemostatic agents. *Successful practice sessions instill confidence in the ability to manage care and provide an opportunity for questions and exploring alternatives.*

Linking NANDA, NIC, and NOC

Linkages between NANDA nursing diagnoses, nursing interventions, and nursing outcomes for the client with hemophilia are illustrated in Chart 34–4.

Community-Based Care

Discuss the following topics when preparing the client with a bleeding disorder and the family for home care:

- Recognizing the manifestations of internal bleeding: pallor, weakness, restlessness, headache, disorientation, pain, swelling. These manifestations require emergency medical care and should be reported immediately.
- Applying cold packs and immobilizing the joint for 24 to 48 hours if hemarthrosis occurs.
- Using analgesics for pain; avoiding prescription and over-the-counter drugs containing aspirin.
- Ensuring a safe home environment (e.g., padding sharp edges of furniture, using transition lighting or a night light; avoiding scatter rugs, and wearing protective gloves when working in the house or yard).
- Using safe grooming practices such as electric razors.
- Wearing a Medic-Alert bracelet in case of accident.
- Practicing good dental hygiene to decrease potential tooth decay and extractions. If dental procedures are necessary, discuss the need for prophylactic factor administration with the dentist and physician.
- Following safer sex practices.
- Preparing and administering intravenous medications.

Refer the client and family to a local hemophilia or bleeding disorders support group. Provide contact information for national organizations and information clearinghouses, such as the National Hemophilia Foundation.

NANDA, NIC, AND NOC LINKAGES
CHART 34–4 **The Client with Hemophilia**

NANDA

Ineffective Health Maintenance

↓

NIC

Health Education

Self-Responsibility Facilitation

Teaching: Procedure/Treatment

↓

NOC

Health-Seeking Behavior

Knowledge: Health Behaviors

Knowledge: Treatment Regimen

Data from *NANDA's Nursing Diagnoses: Definitions & Classification 2005–2006* by NANDA International (2005), Philadelphia; *Nursing Interventions Classification (NIC)* (4th ed.) by J. M. Dochterman & G. M. Bulechek (2004), St. Louis, MO: Mosby; and *Nursing Outcomes Classification (NOC)* (3rd ed.) by S. Moorhead, M. Johnson, and M. Mass (2004), St. Louis, MO: Mosby.

THE CLIENT WITH DISSEMINATED INTRAVASCULAR COAGULATION

Disseminated intravascular coagulation (DIC) is a disruption of hemostasis characterized by widespread intravascular clotting and bleeding. It may be acute and life threatening or relatively mild. DIC is a clinical syndrome that develops as a complication of a wide variety of other disorders (Box 34–8). Sepsis is the most common cause of DIC. Gram-negative and gram-positive bacteria as well as viruses, fungi, and protozoal infections may lead to DIC (McCance & Huether, 2006).

Pathophysiology

DIC is triggered by endothelial damage, release of tissue factors into the circulation, or inappropriate activation of the clotting cascade by an endotoxin. Both the intrinsic and the extrinsic clotting cascade may be activated, although the extrinsic cascade usually is the one activated. Extensive thrombin

BOX 34–8 **Conditions That May Precipitate Disseminated Intravascular Coagulation**

Tissue Damage

- Trauma: burns, gunshot wounds, frostbite, head injury
- Obstetric complications: septic abortion, abruptio placentae, amniotic fluid embolus, retained dead fetus
- Neoplasms: acute leukemia, adenocarcinomas
- Hemolysis
- Fat embolism

Vessel Damage

- Aortic aneurysm
- Acute glomerulonephritis
- Hemolytic uremic syndrome

Infections

- Bacterial infection or sepsis
- Viral or mycotic infections
- Parasitic or rickettsial infection

entering the systemic circulation overwhelms natural anticoagulants, leading to unrestricted clot formation (McCance & Huether, 2006). Clotting may be localized to an individual organ, or widespread with deposition of small thrombi and emboli throughout the microvasculature (Kasper et al., 2005). The widespread clotting consumes clotting factors (prothrombin, platelets, factor V, and factor VIII in particular) and activates fibrinolytic processes with anticoagulant production. As a result, hemorrhage occurs (Figure 34–14 ■).

The sequence of DIC follows:

1. Endothelial damage, tissue factors, or toxins stimulate the clotting cascade.
2. Excess thrombin within the circulation overwhelms naturally occurring anticoagulants.
3. Widespread clotting occurs within the microvasculature.
4. Thrombi and emboli impair tissue perfusion, leading to ischemia, infarction, and necrosis.
5. Clotting factors and platelets are consumed faster than they can be replaced.
6. Clotting activates fibrinolytic processes, which begin to break down clots.
7. Fibrin degradation products (FDPs, potent anticoagulants) are released, contributing to bleeding.
8. Clotting factors are depleted, the ability to form clots is lost, and hemorrhage occurs.

Manifestations

The manifestations of DIC result from both clotting and bleeding, although bleeding is more obvious, especially in acute DIC. Bleeding ranges from oozing blood following an injection to frank hemorrhage from every body orifice (see box below). Chronic DIC may be asymptomatic, or may present with peripheral cyanosis, thrombosis, and pregangrenous changes in the fingers and toes, nose, and genitalia (Kasper et al., 2005).

MANIFESTATIONS of DIC

- Frank hemorrhage from incisions
- Oozing of blood from punctures, intravenous catheter sites
- Purpura, petechiae, bruising
- Cyanosis of extremities
- Gastrointestinal bleeding or hemorrhage
- Dyspnea, tachypnea, bloody sputum
- Tachycardia, hypotension
- Hematuria, oliguria, acute renal failure
- Manifestations of increased intracranial pressure: decreased level of consciousness, papillary, motor, and sensory changes
- Mental status changes

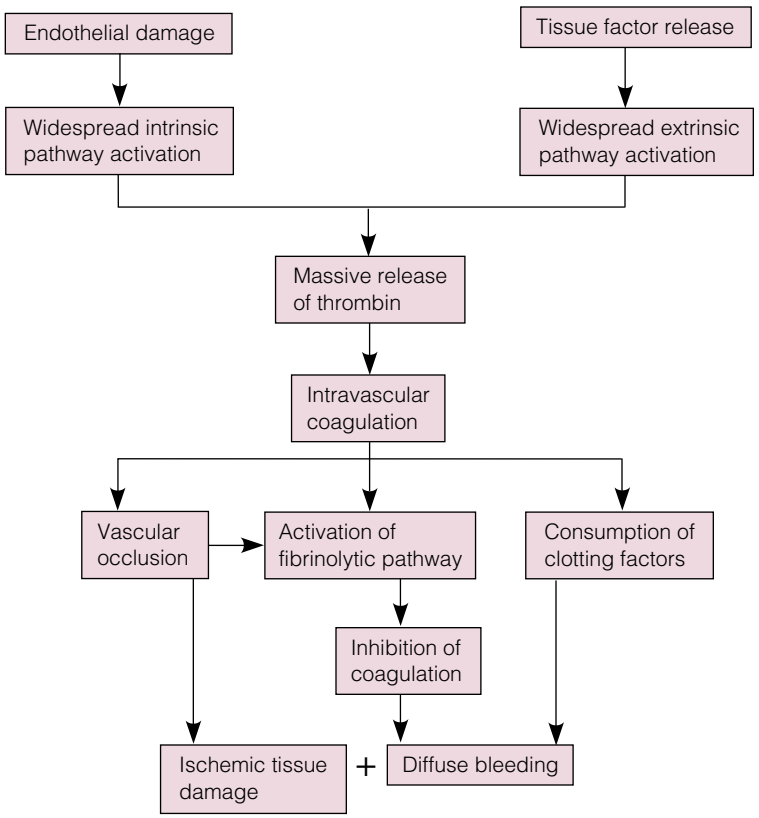


Figure 34–14 ■ Disseminated intravascular coagulation (DIC). Endothelial cell injury or release of tissue factors activate the intrinsic or extrinsic clotting pathway (or both). As a result, numerous microthrombi form throughout the vasculature, causing ischemic tissue damage. Simultaneously, rapid consumption of clotting factors and activation of fibrinolytic mechanisms trigger widespread bleeding.

MEDIALINK Case Study: DIC

INTERDISCIPLINARY CARE

Treatment of DIC is directed toward treating the underlying disorder and preventing further bleeding or massive thrombosis. Treatment stabilizes the client, reduces complications, and allows recovery to occur; it does not cure DIC (Kasper et al., 2005).

Diagnosis

Diagnostic tests are used to confirm the diagnosis of DIC and evaluate the risk for hemorrhage.

- *CBC* and *platelet count* are used to evaluate the hemoglobin, hematocrit, and number of circulating platelets. *Schistocytes,* fragmented RBCs, may be noted due to cell trapping and damage within fibrin thrombi. The platelet count is decreased.
- *Coagulation studies* show prolonged *prothrombin time (PT), partial thromboplastin time (PTT),* and *thrombin time,* and a low *fibrinogen level* due to depletion of clotting factors. The fibrinogen level helps predict bleeding in DIC: As it falls, the risk of bleeding increases (Kasper et al., 2005).
- *Fibrin degradation products (FDPs)* or *fibrin split products (FSPs)* are increased due to the fibrinolysis that occurs with DIC.

Treatments

When bleeding is the major manifestation of DIC, fresh frozen plasma and platelet concentrates are given to restore clotting factors and platelets. Heparin, although controversial, may be administered. Heparin interferes with the clotting cascade and may prevent further clotting factor consumption due to uncontrolled thrombosis. It is used when bleeding is not controlled by plasma and platelets, as well as when the client has manifestations of thrombotic problems such as acrocyanosis and possible gangrene. Long-term heparin therapy (administered by injection or continuous infusion using a portable pump) may be necessary for clients with chronic DIC.

NURSING CARE

Assessment

Nurses can be instrumental in identifying early manifestations of DIC, facilitating timely intervention. Focused nursing assessment for DIC includes:

- *Health history:* Recent abortion (spontaneous or therapeutic) or current pregnancy; presence of a known malignant tumor; history of abnormal bleeding episodes or a hematologic disorder.
- *Physical examination:* Bleeding from puncture wounds (e.g., injections), IV sites, incisions; hematuria, obvious or occult blood in emesis or stool, epistaxis, other abnormal bleeding; vital signs; heart and breath sounds; abdominal assessment including girth, contour, bowel sounds, tenderness or guarding to palpation; color, temperature, skin condition of hands, feet, and digits; petechiae or purpura of skin, mucous membranes.
- *Diagnostic tests:* CBC with hemoglobin, hematocrit; platelet count; coagulation studies; evaluations of organ system function (e.g., liver and renal function tests); CT scans of the head and abdomen.

Nursing Diagnoses and Interventions

Clients with acute DIC often are critically ill, with multiple nursing care needs. Priority nursing diagnoses discussed in this section focus on impaired tissue perfusion and gas exchange, pain, and fear. Septic shock may precipitate DIC; hemorrhagic shock may occur as a complication of DIC. See Chapter 11 ∞ for nursing diagnoses and interventions related to these problems.

Ineffective Tissue Perfusion

Thrombi and emboli forming throughout the microcirculation affect the perfusion of multiple organs and tissues. Additionally, bleeding due to clotting factor consumption affects cardiac output and blood flow to these tissues.

- Assess extremity pulses, warmth, and capillary refill. Monitor level of consciousness (LOC) and mental status. *Monitoring central and peripheral tissue perfusion facilitates early treatment of impaired perfusion.*

PRACTICE ALERT

Promptly report complaints of chest pain, changes in mental status, LOC, tissue perfusion, respirations, gastrointestinal function, and urinary output. Chest pain or respiratory changes (tachypnea, dyspnea, orthopnea) may be due to angina, pulmonary embolism, or bleeding into lung tissue. Changes in mentation or LOC can indicate cerebral ischemia. A painful, pale, and cold extremity with no or diminished pulses indicates arterial occlusion. Prompt intervention is critical to save the extremity. Acute abdominal pain, decreased bowel sounds, and GI bleeding may indicate mesenteric occlusion, a surgical emergency. Decreased urine output may signify renal artery thrombosis; renal failure may develop.

- Carefully reposition at least every 2 hours. *Position changes facilitate circulation and tissue perfusion and also provide an opportunity to assess for purpura, pallor, and bleeding.*
- Discourage crossing the legs, and do not elevate the knees on the bed or with a pillow. *These positions may impair arterial and venous flow to the lower legs and feet, increasing vascular stasis and the risk for thrombosis.*
- Minimize use of tape on the skin, using binders, nonadhesive dressings, and other devices as needed. *Preventing skin trauma reduces the risk for bleeding and potential infection.*

Impaired Gas Exchange

Microclots in the pulmonary vasculature are likely to interfere with gas exchange in the client with DIC.

- Monitor oxygen saturation continuously. Administer oxygen as ordered. *Oxygen saturation levels are a noninvasive means of assessing gas exchange. Supplemental oxygen promotes gas exchange and reduces cardiac work, relieving dyspnea.*

PRACTICE ALERT
Monitor arterial blood gas results; report abnormal results to the physician. Low Pa_{O_2} and rising Pa_{CO_2} levels indicate impaired gas exchange and may signify the need for additional treatment.

- Place in Fowler's or high-Fowler's position as tolerated. *Elevating the head of the bed improves diaphragmatic excursion and alveolar ventilation.*
- Maintain bed rest. *Bed rest reduces oxygen demands and cardiac work.*
- Encourage deep breathing and effective coughing. *Increased respiratory depth and clearance of secretions from airways improves alveolar ventilation and oxygenation.*
- Cautious nasotracheal suctioning may be instituted if cough is ineffective or an endotracheal tube is in place. *Removal of secretions facilitates ventilation and oxygenation. However, care must be used to minimize suction-induced hypoxia and airway trauma.*
- Administer analgesics and antianxiety drugs as needed to control pain and anxiety. Provide reassurance and comfort measures. *Pain and anxiety increase the respiratory rate and decrease the depth of respirations, reducing effective ventilation and gas exchange.*

Pain

Both the underlying cause of DIC and tissue ischemia from microvascular clots can cause pain. Identifying the etiology of pain is important to identify potential complications or harmful effects of DIC and to institute effective treatment.

- Use a standard pain scale to evaluate and monitor pain and analgesic effectiveness. *Monitoring pain and response to medication facilitates development of an appropriate and effective treatment plan.*

PRACTICE ALERT
Notify the physician promptly of new or a sudden increase in pain, especially when accompanied by changes in assessment findings. New or increased complaints of pain may signify increased circulatory impairment and ischemic changes in tissues such as the heart, bowel, or extremities. Circulation to a painful, pale or cyanotic, or cold extremity may be occluded by an arterial clot. Prompt intervention is necessary to save the extremity. Acute abdominal pain may signify mesenteric occlusion, a surgical emergency. Anginal pain may indicate occlusion of coronary arteries.

- Handle extremities gently. *Gentle handling reduces the risk of further injury to and pain in ischemic tissues.*
- Apply cool compresses to painful joints. *Application of cold decreases pain through the gate-control mechanism, inhibiting the dorsal horn of the spinal cord and reducing the sensation of pain.*

PRACTICE ALERT
Continuously monitor effects of analgesics, mental and respiratory status. Analgesics may mask manifestations of neurologic impairment due to thromboembolism, and may depress the respiratory center, further impairing gas exchange. Judicious analgesic administration with careful monitoring is vital to safely provide effective pain relief.

Fear

The underlying serious illness and a complication such as DIC result in an uncertain prognosis, often accompanied by fear.

- Encourage the client and family to verbalize concerns. *This helps the client and family identify their concerns and frame questions.*
- Answer questions truthfully. *Providing honest answers is vital to developing a therapeutic nurse–client relationship. Accurate responses allow the client and family to set priorities as they plan for an uncertain future.*
- Help the client and family identify coping strategies to manage this significant situational stressor. *Implementing past effective coping methods may provide the skills to manage the current crisis.*
- Provide emotional support. *The presence of a caring nurse helps reduce the fear and anxiety associated with a crisis.*
- Maintain a calm environment. *A calm environment provides reassurance that the situation is in control, reduces anxiety, and promotes rest.*
- Respond promptly when the client calls for help. *Prompt response to expressed needs helps develop a trusting relationship and a sense of security that assistance is readily available.*
- Teach relaxation techniques. *Relaxation techniques can reduce muscle tension and other signs of anxiety. Gaining control over physical responses can help the client gain a sense of control over the situation.*

Community-Based Care

Although the immediate crisis of acute DIC is resolved prior to discharge, the client may have some continuing effects of the disorder, such as impaired tissue integrity of distal extremities. Teach the client and family about specific care needs, such as foot care (see Box 35–4 ∞) or dressing changes. Provide instruction about any continuing medications and follow-up care.

Clients with chronic DIC may require continuing heparin therapy, using either intermittent subcutaneous injections or a portable infusion pump. Teach the client and family members how to administer the injection or manage the infusion pump. Provide a referral to home health care or a home intravenous management service for assistance. Discuss the manifestations of excessive bleeding or recurrent clotting that need to be reported to the physician.

EXPLORE MEDIALINK

Prentice Hall Nursing MediaLink DVD-ROM

Audio Glossary
NCLEX-RN® Review

Animation/Video

Sickle Cell Anemia

COMPANION WEBSITE www.prenhall.com/lemone

Audio Glossary
NCLEX-RN® Review
Care Plan Activity: Acute Myelocytic Leukemia
Case Studies
- *Disseminated Intravascular Coagulation*
- *Immune Thrombocytopenic Purpura*

MediaLink Applications
- *Blood Alternatives*
- *Sickle Cell Anemia*
- *Stem Cell Transplant*
- *Synthetic Blood Products*

Links to Resources

CHAPTER HIGHLIGHTS

- Anemia is the most common disorder of the red blood cells; nutritional deficiencies are the most common causes of anemia. Its manifestations relate to the function of RBCs and hemoglobin, transporting oxygen to the cells: fatigue, increased respiratory and heart rates, shortness of breath with activity, and pallor.
- Genetically transmitted disorders such as sickle cell disease and thalassemia can cause significant anemia and associated problems in affected populations. These clients require teaching and episodic acute care for crises such as vaso-occlusive crisis in sickle cell disease.
- Nursing care related to anemia is primarily educational to prepare the client for effective self-care, including diet, prescribed medications, and measures to prevent sickling episodes (for clients with sickle cell disease).
- Leukemia and lymphomas are the primary disorders of white blood cells and lymphoid tissues.
- Manifestations of the leukemias reflect the altered ability of abnormal WBCs to perform effective immune surveillance and crowding of the bone marrow and other organs by rapidly proliferating cells. Frequent sore throats, increased risk for infection, and manifestations of anemia and thrombocytopenia are seen, as well as an enlarged spleen and abdominal pain.
- Four major subgroups of leukemia are identified, acute and chronic myeloid leukemias, and acute and chronic lymphocytic (or lymphoblastic) leukemias. The primary population affected differs for each of these leukemias, as does their course.
- Genetic alterations and certain viruses are linked to the development of leukemia, as are exposure to chemotherapy drugs, environmental toxins, and ionizing radiation.
- Lymphocytic leukemias and lymphomas are closely related disorders.
- Nursing care for clients with leukemia and lymphoma focuses on reducing the risk for infection and bleeding, managing the effects of chemotherapy and radiation therapy, and, in some cases, caring for clients before and after bone marrow or stem cell transplant.
- The major risks associated with bone marrow and stem cell transplant are infection prior to and immediately following the transplant and graft-versus-host disease, a potentially fatal condition. A pruritic rash and desquamation of the palms and soles; abdominal pain, nausea, and diarrhea; and jaundice and elevated liver enzymes are common early manifestations of GVHD.
- The treatment of and nursing care for clients with lymphomas (including Hodgkin's lymphoma and non-Hodgkin's lymphomas) is similar to that provided for clients with leukemia.
- Multiple myeloma is a malignancy of plasma cells, B lymphocytes that produce antibodies. Circulating M proteins and Bence Jones proteins in the urine are seen in multiple myeloma. The usual presenting manifestation is bone pain. Pathologic fractures and hypercalcemia are common complications of multiple myeloma as bone is destroyed.
- Bleeding and clotting disorders can result from either inadequate platelets (thrombocytopenia) or disruption of the clotting mechanisms (hemophilia, DIC). Petechiae and purpura are common manifestations of bleeding/clotting disorders.
- Hemophilias are genetically transmitted disorders. Hemophilia A and B are transmitted on the X chromosome (sex-linked) from mother to son. Von Willebrand's disease, the most common bleeding disorder, is transmitted as an autosomal dominant disorder and affects men and women equally.
- Hemophilias are treated by replacement of the missing clotting factor and measures to prevent injury and bleeding.
- Disseminated intravascular coagulation is a disorder of widespread microvascular clotting. It commonly is precipitated by sepsis, but also may occur with conditions such as major trauma, malignancy, or as an obstetric emergency.
- In DIC, platelets and clotting factors are consumed by the abnormal clotting processes, leading the manifestations of bleeding, including frank hemorrhage, hematuria, oozing blood from parenteral and intravenous injection sites, and GI bleeding. Blood flow to organs and tissues is compromised by clot

formation, leading to manifestations such as cyanosis of extremities, abdominal pain, renal failure, and changes in mental status and level of consciousness. Nursing care is supportive, focusing on administering prescribed treatments and monitoring and supporting cardiovascular, respiratory, and renal function.

TEST YOURSELF NCLEX-RN® REVIEW

1 In assessing a female client with moderate anemia, the nurse would expect to find which of the following?
   1. hematocrit 45%
   2. pulse rate 140
   3. complaints of shortness of breath with exercise
   4. WBC 14,000/μL

2 The nurse following a client after gastric resection observes carefully for evidence of nutritional deficiency anemia related to malabsorption, including:
   1. numbness and tingling of extremities.
   2. steatorrhea.
   3. dark yellow or bronze skin color.
   4. bone pain.

3 Which of the following nursing diagnoses would be of highest priority for the client hospitalized for a bone marrow transplant to treat relapse of acute myelocytic leukemia?
   1. *Disturbed Body Image*
   2. *Ineffective Protection*
   3. *Anxiety*
   4. *Imbalanced Nutrition: Less than Body Requirements*

4 The nurse caring for a client with acute myeloid leukemia plans which of the following nursing interventions during hospitalization? (Select all that apply.)
   1. Place in a private room.
   2. Implement airborne infection control precautions.
   3. Assist with oral hygiene after meals.
   4. Monitor rectal temperature q4h.
   5. Request soft, bland diet.

5 A client with non-Hodgkin's lymphoma tells the nurse, "I might as well give up on dating. No woman will want me now." What is the most appropriate response?
   1. "It sounds like you are concerned about the effects of this disease and the proposed treatment plan."
   2. "Don't worry. Malignant lymphomas are very treatable when caught in an early stage of the disease."
   3. "Well, you may never be able to have children all right, but there are other ways to have a satisfying relationship with a woman."
   4. "Lots of women find bald men attractive; besides, your hair may grow back soft and curly."

6 The nurse caring for a client with lymphoma who is being started on the CHOP chemotherapy regimen understands that chemotherapy drugs are used in combination to:
   1. target malignant cells in different organs.
   2. prevent the development of adverse effects.
   3. target different phases of the cell cycle.
   4. support growth and development of normal cells.

7 A client with multiple myeloma calls the home health nurse complaining of severe back pain of new onset. The appropriate response by the nurse is to:
   1. reassure the client that bone pain is expected with this disease.
   2. inquire about the client's use of NSAIDs and analgesics to manage pain.
   3. suggest use of a back brace to reduce pain.
   4. notify the physician of the onset of new pain.

8 The nurse observes reddish-purple spots and areas of purple bruising on a newly admitted client. Which laboratory results support this assessment finding?
   1. hematocrit 28%
   2. platelets 6000/mm^3
   3. INR 4.0
   4. WBC 4500/mm^3

9 A client whose husband has hemophilia asks if her newborn baby girl could have the disease. The nurse's response is based on the knowledge that:
   1. the most common forms of hemophilia are transmitted as sex-linked recessive disorders; her daughter is at risk for carrying the defective gene.
   2. because hemophilia is a sex-linked recessive disorder carried on the Y chromosome, her daughter has no risk of having or carrying the disease.
   3. hemophilia is an autosomal dominant disorder; therefore, her daughter has a 50% chance of having the disorder.
   4. although hemophilia is genetically transmitted, its pattern of inheritance is unknown, and her daughter will need to be tested for the defective gene.

10 The nurse administering platelets to a client with disseminated intravascular coagulation (DIC) understands that the intended effect of this treatment is to:
   1. replace specific clotting factors.
   2. promote intravascular clotting.
   3. restore tissue oxygenation.
   4. replace depleted platelets.

See Test Yourself answers in Appendix C.

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