



Chapter 27

The Child with a Condition of the
Blood, Blood-Forming Organs, or
Lymphatic System



Objectives

- Summarize the components of blood.
- Recall normal blood values of infants and children.
- List two laboratory procedures commonly performed on children with blood disorders.
- Review the effects of severe anemia on the heart.
- Compare and contrast four manifestations of bleeding into the skin.



Objectives (*cont.*)

- List the symptoms, prevention, and treatment of iron-deficiency anemia.
- Recommend four food sources of iron for a child with iron-deficiency anemia.
- Examine the pathology and signs and symptoms of sickle cell disease.
- Describe four types of sickle cell crisis.
- Devise a nursing care plan for a child with sickle cell disease.



Objectives (*cont.*)

- Recognize the effects on the bone marrow of increased red blood cell production caused by thalassemia.
- Recall the pathology and signs and symptoms of hemophilia A and hemophilia B.
- Identify the nursing interventions necessary to prevent hemarthrosis in a child with hemophilia.



Objectives (*cont.*)

- Plan the nursing care of a child with leukemia.
- Discuss the nursing care of a child receiving a blood transfusion.
- Discuss the effects of chronic illness on the growth and development of children.
- Recall the stages of dying.



Objectives (*cont.*)

- Contrast age-appropriate responses to a sibling's death and the nursing interventions required.
- Formulate techniques the nurse can use to facilitate the grieving process.
- Discuss the nurse's role in helping families to deal with the death of a child.

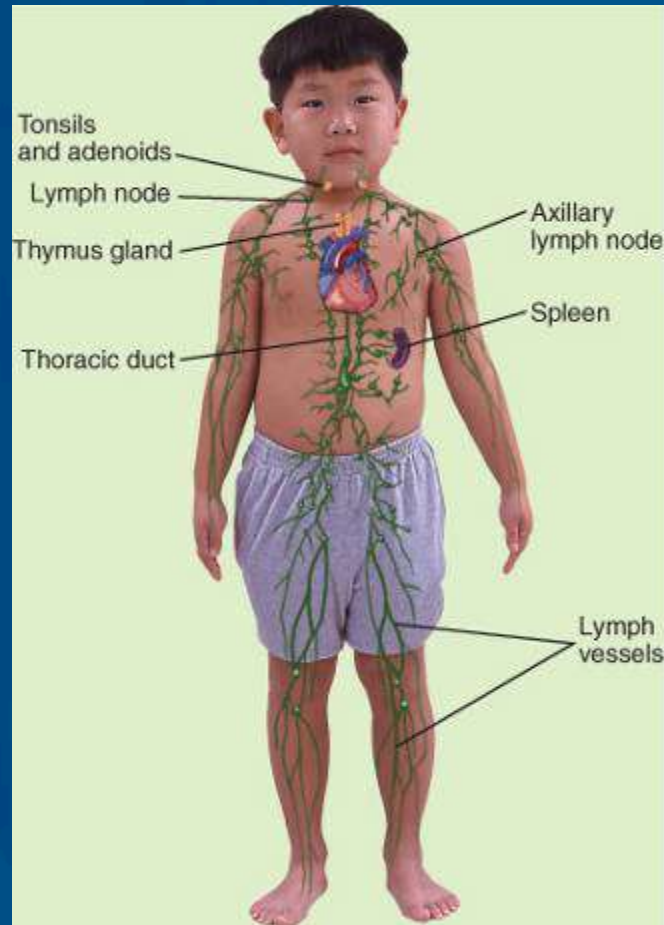


Blood Dyscrasias

- Occur when blood values exceed or fail to form correctly or fail to meet normal standards
- During childhood, RBCs are formed in the marrow of the long bones; by adolescence, *hematopoiesis* takes place in the marrow of the ribs, sternum, vertebrae, pelvis, skull, clavicle, and bone marrow
- RBC production is regulated by erythropoietin
 - Substance is produced by the liver of the fetus
 - At birth, the kidneys take over this process



Lymphatic System



LYMPHATIC SYSTEM

- The increased size of tonsils and adenoids is normal in preschool and school-age children and is one of the body's defense mechanisms.
- The thymus gland is important in the development of the immune response in newborns.
- Preterm and term infants are at greater risk for viral and bacterial infections because of immature T-cell activity.



Lymphatic System (*cont.*)

- Drains regions of the body to lymph node
 - Where infectious organisms are destroyed
 - Antibody production is stimulated
- Lymphadenopathy is an enlargement of lymph nodes
 - Indicative of infection or disease



Lymphatic System (*cont.*)

- Spleen is largest organ of the lymphatic system
- One of the main functions is to bring blood into contact with lymphocytes
- Most common pathological condition is enlargement (splenomegaly)
- Enlarges during infections, congenital and acquired hemolytic anemias, and liver malfunction



Circulating Blood

- Consists of two portions
- Plasma
- Formed elements
 - Erythrocytes
 - Leukocytes (white blood cells [WBCs])
 - Thrombocytes (platelets)
- Erythrocytes
 - Transport oxygen and carbon dioxide to and from the lungs and tissues
- Leukocytes act as the body's defense against infection
- Lymphocytes are produced in the lymphoid tissues of the body



Anemias

- Can result from many different underlying causes
 - A reduction in the amount of circulating hemoglobin (Hgb) reduces the oxygen-carrying ability of the blood
 - An Hgb below 8 g/dl results in an increased cardiac output and a shunting of blood from the periphery to the vital organs
 - Can result in pallor, weakness, tachypnea, SOB, CHF



Iron-Deficiency Anemia

- Most common nutritional deficiency of children
 - Incidence is highest during infancy (from 9th to 24th month) and adolescence
- May be caused by severe hemorrhage, inability to absorb iron received, excessive growth requirements, or an inadequate diet
 - Giving whole cow's milk to infants can lead to GI bleeding, leading to anemia



Iron-Deficiency Anemia (*cont.*)

- Manifestations
 - Pallor
 - Irritability
 - Anorexia
 - Decrease in activity
 - Infants may be overweight due to excessive milk consumption
- Blood tests
 - RBC count
 - Hgb and hematocrit
 - Morphological cell changes
 - Iron concentrations
- Stool may be tested for occult blood



Iron-Deficiency Anemia (*cont.*)

- Untreated, iron-deficiency anemia will progress slowly
 - In severe cases, heart muscle becomes too weak to function
 - Children with long-standing anemia may also show growth retardation and cognitive changes
- Treatment
 - Iron, usually ferrous sulfate, orally 2 to 3 times a day
 - Vitamin C aids in absorption



Nursing Tip

- Oral iron supplements should not be given with milk or milk products because milk interferes with iron absorption



Parent Education

- Nurse stresses importance of breastfeeding for the first 6 months and the use of iron-fortified formula throughout the first year of life
- Stools of infants taking oral iron supplements are tarry green
- Do not give iron with milk
- To increase absorption, give the iron between meals when digestive acid concentration is at its highest

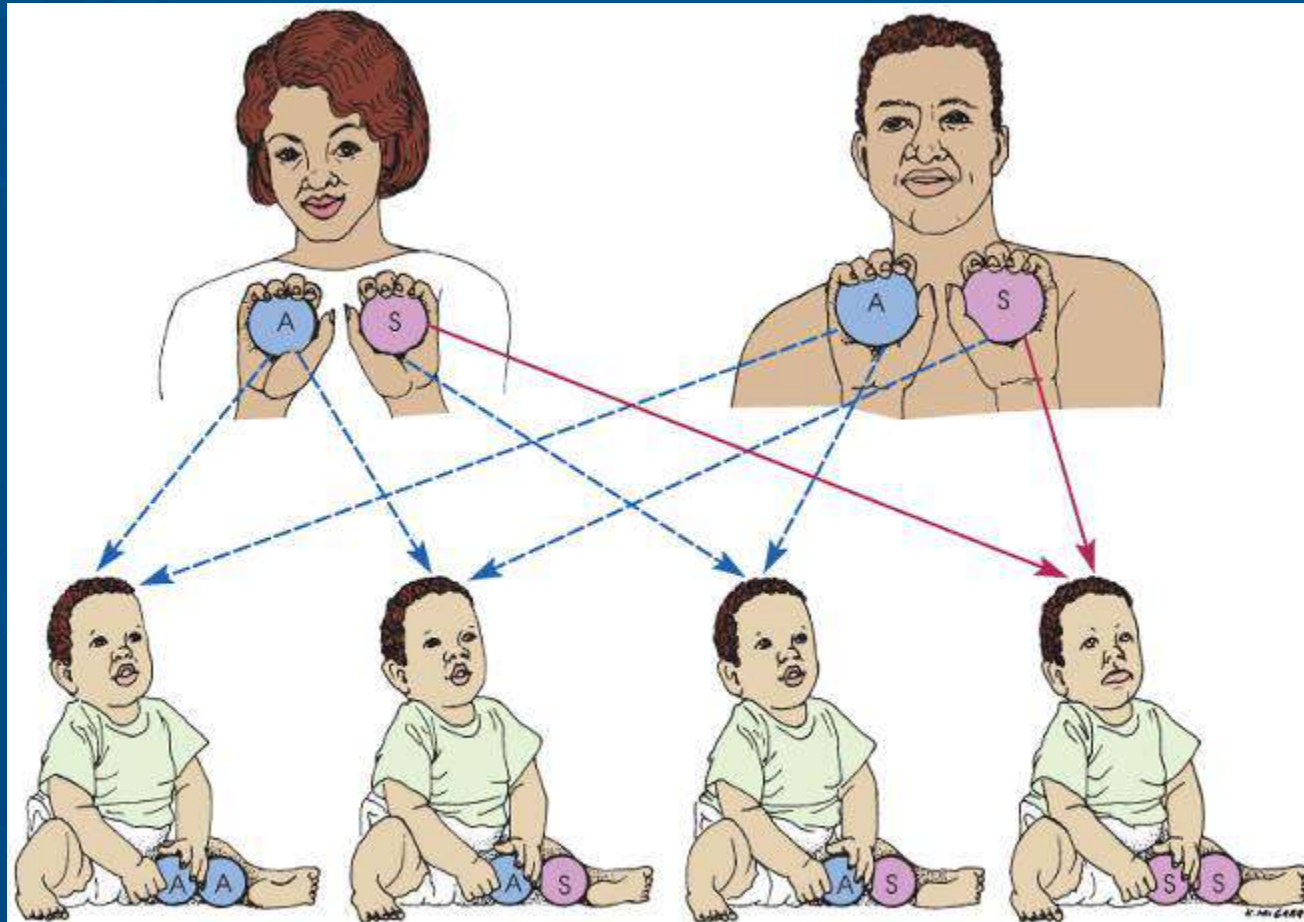


Sickle Cell Disease

- Inherited defect in the formation of hemoglobin
- Sickling (clumping) caused by decreased blood oxygen levels may be triggered by dehydration, infection, physical or emotional stress, or exposure to cold
 - Membranes of these cells are fragile and easily destroyed
 - Their crescent shape makes it difficult for them to pass through the capillaries, causing a pileup of cells in the small vessels
 - May lead to thrombosis, can be very painful
- Hemosiderosis (iron deposits into body organs) is a complication of the disease



Transmission of Sickle Cell Disease from Parents to Children





Two Types of Sickle Cell Disease

- Sickle cell trait (asymptomatic)
 - Blood of the patient contains a mixture of Hgb A and sickle (Hgb S)
 - Proportions of Hgb S are low because the disease is inherited from only one parent
- Hgb and RBC counts are normal
- Sickle cell anemia (more severe)
- Clinical symptoms do not appear until the last part of the first year of life
 - May be an unusual swelling of the fingers and toes
 - Symptoms caused by enlarging bone marrow sites that impair circulation to the bone and the abnormal sickle cell shape that causes clumping, obstruction in the vessel, and ischemia to the organ the vessel supplies



Manifestations

- Hgb level ranges 6 to 9 g/dL or lower
 - Child is pale, tires easily, and has little appetite
- Sickle cell crises are painful and can be fatal
 - Symptoms: severe abdominal pain, muscle spasms, leg pain, or painful swollen joints may be seen
 - Fever, vomiting, hematuria, convulsions, stiff neck, coma, or paralysis can result
 - Risk for stroke as a complication of a vaso-occlusive sickle cell crisis



Types of Sickle Cell Crises

- Vaso-occlusive (painful crises)
- Splenic sequestration
- Aplastic crises
- Hyperhemolytic



Health Promotion

- During sickle cell crisis, anticipate the child's need for tissue oxygenation, hydration, rest, protection from infection, pain control, blood transfusion, and emotional support for this life-threatening illness



Therapies and Goals

- Erythropoietin and some chemotherapy regimens can increase the production of fetal Hgb and reduce complications
- Routine splenectomy is *not* recommended because the spleen generally atrophies on its own because of fibrotic changes that take place in patients with sickle cell disease
- Prevent infection, dehydration, hypoxia, and sickling



Thalassemia

- Group of hereditary blood disorders in which the patient's body cannot produce sufficient adult Hgb
- RBCs are abnormal in size and shape and are rapidly destroyed; results in chronic anemia
- Body attempts to compensate by producing large amounts of fetal Hgb



Thalassemia (*cont.*)

- Categorized according to the polypeptide chain affected
 - *Beta-thalassemia* is the most common variety; involves impaired production of beta chains
 - Two forms
 - Thalassemia minor
 - Thalassemia major, also known as *Cooley's anemia*
 - Can also occur from spontaneous mutations



Thalassemia Minor

- Also termed beta-thalassemia trait, occurs when the child inherits a gene from only one parent
 - Heterozygous inheritance
- Associated with mild anemia
- Often misdiagnosed as having iron-deficiency anemia
- Symptoms minimal
 - Pale
 - Possible splenomegaly
- May lead a normal life with the illness going undetected
- Of genetic importance, particularly if both parents are carriers of the trait



Thalassemia Major (Cooley's Anemia)

- Child is born with a more serious form of the disease when two thalassemia genes are inherited (homozygous inheritance)
- Progressive, severe anemia
- Evident within the second 6 months of life
- Child is pale, hypoxic, poor appetite, and may have a fever



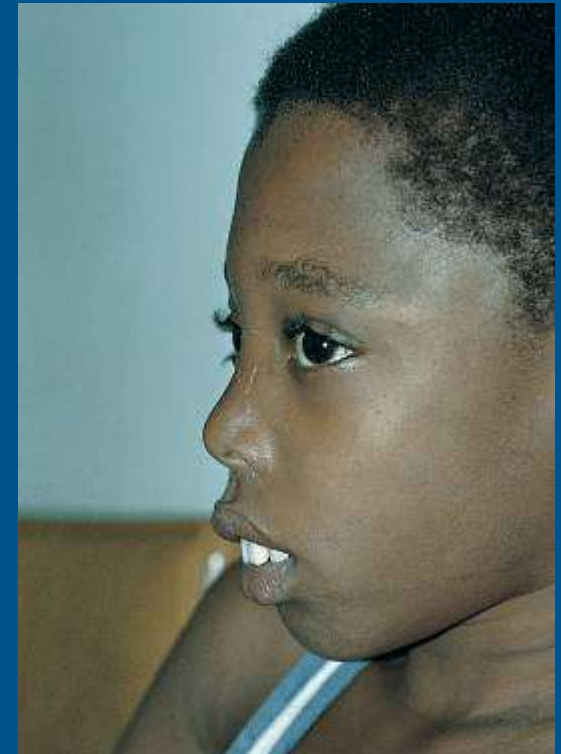
Thalassemia Major (Cooley's Anemia) (*cont.*)

- Jaundice that progresses to a muddy bronze color resulting from hemosiderosis
- Liver enlarges and the spleen grows enormously
- Abdominal distention is great
- Increases pressure on the chest organs
- Cardiac failure caused by profound anemia is a constant threat



Thalassemia Major (Cooley's Anemia) (*cont.*)

- Bone marrow space enlarges to compensate for an increased production of blood cells
 - Hematopoietic defects and a massive expansion of the bone marrow in the face and skull result in changes in the facial contour
 - Teeth protrude due to an overgrowth of the upper jawbone
 - Bone becomes thin and is subject to fracture





Thalassemia Major (Cooley's Anemia) (*cont.*)

- Diagnosis
 - Family history of thalassemia
 - Radiographic bone growth studies
 - Blood test
 - Hemoglobin electrophoresis is helpful in diagnosing type and severity



Thalassemia Major (Cooley's Anemia) (*cont.*)

- Goals of therapy
 - Maintain hemoglobin levels to prevent overgrowth of bone marrow and resultant deformities
 - Provide for normal growth and development and physical activity
 - Prevention or early treatment of infection is important
 - Some may require a splenectomy due to degree of splenomegaly



Thalassemia Major (Cooley's Anemia) (*cont.*)

- Mainstay of treatment
 - Frequent blood transfusions to maintain Hgb above 10 g/dL
 - Because of the number of transfusions, hemosiderosis is seen in the spleen, liver, heart, pancreas, and lymph glands
 - Deferoxamine mesylate (Desferal), an iron-chelating agent is given to counteract this side effect
 - A splenectomy may be needed to increase comfort, increase ability to move about, and to allow for more normal growth



Thalassemia Major (Cooley's Anemia) (*cont.*)

- Nursing measures
 - Adhere to the principles of long-term care
 - Whenever possible, have the same nurse assigned to the child
 - Observing the patient during blood transfusions for any adverse reactions
 - Monitoring vital signs
 - Providing for the emotional health of the child and family is essential



Bleeding Disorders



Hemophilia

- One of the oldest hereditary diseases known to man
- Blood does not clot normally
- Congenital disorder confined almost exclusively to males
 - Is transmitted by symptom-free females



Hemophilia (*cont.*)

- Inherited sex-linked recessive trait
 - Defective gene is located on the X, or female, chromosome
 - Fetal blood samples detect hemophilia
- Two most common types
 - *Hemophilia B* (Christmas disease [a factor IX deficiency])
 - *Hemophilia A* (a deficiency in factor VIII)
- A deficiency in any one of the factors will interfere with normal blood clotting



Hemophilia A

- Caused by a deficiency of coagulation factor VIII, or antihemophilic globulin (AHG)
- Severity dependent on level of factor VIII in the plasma
- Some patients' lives can be endangered by a minor scratch, while others may simply bruise more easily than the average person
- Aim of therapy is to increase level of factor VIII to ensure clotting
- This is checked by a blood test call *partial thromboplastin time (PTT)*



Manifestations of Hemophilia

- Can be diagnosed at birth because factor VIII cannot cross the placenta and be transferred to the fetus
 - Usually not apparent in the newborn unless abnormal bleeding occurs at the umbilical cord or after circumcision
- Normal blood clots in 3 to 6 minutes
 - In severe hemophilia, it can take up to 1 hour or longer



Manifestations of Hemophilia (*cont.*)

- Anemia, leukocytosis, moderate increase in platelets may be seen in hemorrhaging; may also be signs of shock
- Spontaneous hematuria is seen
- Death can result from excessive bleeding, especially if it occurs in the brain or neck
- Severe headache, vomiting, and disorientation may be symptoms



Diagnosis

- Circumstances leading to diagnosis
 - Nosebleed that will not stop
 - Loss of a deciduous tooth
 - Hematomas develop at the injection site of an immunization
 - Hemorrhage into the joint cavity (considered a classic symptom)
- A classic symptom of hemophilia is bleeding into the joints (hemarthrosis)



Treatment of Hemophilia

- If family history exists, a newborn may have certain procedures delayed to prevent bleeding and tissue injury
- Principal therapy is to prevent bleeding by replacing the missing factor
- Recombinant antihemophilic factor, a synthetic product, has eliminated the need for repeated blood transfusions
- Desmopressin acetate (DDAVP) is a nasal spray that can stop bleeding
- Prophylactic care must be provided prior to planned invasive procedures



Treatment of Hemophilia (*cont.*)

- Multidisciplinary approach to assist families to develop healthy coping strategies to deal with a child with a chronic illness
- Difficult for parents not to be overprotective
- The struggle to protect these children and still foster independence and a sense of autonomy is important therefore; allowing the child to participate in decision-making about their care and focusing on their strengths are helpful



Safety Alert

- Drugs that contain salicylates are contraindicated for children with hemophilia



Platelet Disorders

- Reduction or destruction of platelets in the body interferes with the clotting mechanisms
- Skin lesions common to this type of disorder
 - Petechiae
 - Purpura
 - Ecchymosis
 - Hematoma



Idiopathic (Immunological) Thrombocytopenic Purpura (ITP)

- Acquired platelet disorder that occurs in childhood
- Most common of the purpuras
- Cause is unknown but is thought to be an autoimmune reaction to a virus
- Platelets become coated with antiplatelet antibody, seen as “foreign” and are eventually destroyed by the spleen
- ITP occurs in all age groups, with main incidence between 2 and 4 years of age



Manifestations of ITP

- Classic symptom is easy bruising
 - Results in petechiae and purpura
- May have recent history of rubella, rubeola, or viral respiratory infection
 - Interval between exposure and onset is about 2 weeks
- Platelet count below $20,000/\text{mm}^3$ (normal range is between $150,000$ and $400,000/\text{mm}^3$)
 - Diagnosis confirmed by bone marrow aspiration



Treatment of ITP

- Neurological assessments are a priority of care
- Treatment is not indicated in most cases
- If indicated, prednisone, IV gamma globulin, and anti-D antibody are some of the treatment options
- In cases of chronic ITP, a splenectomy may be required
- Drugs to avoid
 - Aspirin
 - Phenylbutazone
 - Phenacetin
 - Caffeine
- Activity is limited during acute states to avoid bruising
- Platelets are usually not given because they are destroyed by the disease process



Complications of ITP

- Bleeding from the GI tract
- Hemarthrosis
- Intracranial hemorrhage
- Prevention may be helped by immunizing all children against the viral diseases of childhood



Disorders of White Blood Cells



Risk for Development of Cancer

- Genetic and environmental factors play a role
- Exposure of the fetus to diagnostic X-rays or therapeutic irradiation for brain tumors, the use of fluoroscopy, ultraviolet (sun) exposure, and some drugs have been associated with the increase in cancer



Leukemia

- Most common form of cancer in childhood
- Refers to a group of malignant diseases of the bone marrow and lymphatic system
- Classified according to what type of WBC affected
- Two most common
 - Acute lymphoid leukemia (ALL)
 - Acute non-lymphoid (myelogenous) leukemia (AMLL or AML)
 - Cytochemical markers, chromosome studies, and immunological markers differentiate the two types



Leukemia (*cont.*)

- A malignant disease of the blood-forming organs that results in an uncontrolled growth of immature WBCs
- Involves a disruption of bone marrow function caused by the overproduction of immature WBCs in the marrow
 - These immature WBCs take over the centers that are designed to form RBCs, and anemia results
- Platelet counts are also reduced
- Invasion of the bone marrow causes weakening of the bone, and pathological fractures can occur



Leukemia (*cont.*)

- Leukemia cells can infiltrate the spleen, liver, and lymph glands, resulting in fibrosis and diminished function
- Cancerous cells invade the CNS and other organs
 - Drain the nutrients
 - Lead to metabolic starvation of the body



Manifestations of Leukemia

- Most common symptoms
 - Initial phase
 - Low-grade fever
 - Pallor
 - Bruising tendency
 - Leg and joint pain
 - Listlessness
 - Abdominal pain
 - Enlargement of lymph nodes
 - Anemia severe despite transfusions
- Gradual or sudden onset
- As it progresses, the liver and spleen become enlarged
- Skin may have a lemon-yellow color
- Petechiae and purpura may be early objective symptoms
- Anorexia, vomiting, weight loss, and dyspnea are also common



Manifestations of Leukemia (*cont.*)

- WBCs not functioning normally, increases risk of infection
- Ulcerations develop around the mucous membranes of the mouth and anal regional
 - Gums tend to bleed





Diagnosis

- Based on history and symptoms
- Results of extensive blood tests
 - Demonstrate presence of leukemic blast cells in the blood, bone marrow, or their tissues
- X-rays of the long bones show changes
- Spinal tap may be done to check for CNS involvement
- Kidney and liver function studies are done
 - The adequacy of their function is essential to the outcomes of chemotherapy



Treatment of Leukemia

- Long-term care given whenever possible in an outpatient setting
- Bone marrow suppression in chemotherapy requires family teaching for infection prevention
- Adequate hydration to minimize kidney damage
- Active routine immunizations must be delayed while receiving immunosuppressive drugs
- Nausea and vomiting are common side effects of chemotherapy; can lead to decreased appetite, weight loss, and generalized weakness
- Meticulous oral care is necessary



Treatment of Leukemia (*cont.*)

- Components of chemotherapy include
 - Induction period
 - Central nervous system prophylaxis for high-risk patients
 - Maintenance
 - Reinduction therapy (if relapse occurs)
 - Extramedullary disease therapy
- Bone marrow transplant



Side Effects of Chemotherapy

- Steroids can mask signs of infection, cause fluid retention, induce personality changes, and cause the child's face to appear moon-shaped
- Certain chemotherapy agents can cause nausea, diarrhea, rash, hair loss, fever, anuria, anemia, and bone marrow depression
- Peripheral neuropathy may be signaled by severe constipation caused by decreased nerve sensations to the bowel



Nursing Care of the Child with Leukemia

- Encourage the child to verbalize feelings
 - Giving permission to discuss their concerns will help clear up misconceptions and to decrease feelings of isolation
- Frequently observe child for infection
- Monitor vital signs and for symptoms of thrombocytopenic bleeding (a common complication of leukemia)
- Meticulous mouth and skin care



Child Receiving a Blood Transfusion

- Hemolytic reactions caused by mismatched blood are rare
- Blood is slowly infused through blood filter to avoid impurities
- Medications are **never** added to blood
- Monitor the child for signs of transfusion reaction (most occur within the first 10 minutes of the transfusion)
- Circulatory overload is a danger in children



Safety Alerts

- If a blood transfusion reaction occurs, stop the infusion, keep the vein open with normal saline solution, and notify the charge nurse
- Take the patient's vital signs and observe closely



Hodgkin's Disease

- A malignancy of the lymph system that primarily involves the lymph nodes
 - May metastasize to the spleen, liver, bone marrow, lungs, or other parts of the body
- Presence of giant multinucleated cells called *Reed-Sternberg cells* is diagnostic of the disease
- Rarely seen before 5 years of age, incidence increases during adolescence and early adulthood
 - Twice as common in boys as in girls



Manifestations of Hodgkin's Disease

- A painless lump along the neck
- Few other manifestations
- More advanced cases, may be unexplained low-grade fever, anorexia, unexplained weight loss, night sweats, general malaise, rash, and itching



Criteria for Staging Hodgkin's Disease

Stage	Criteria
I	Restricted to single site or localized in a group of lymph nodes; asymptomatic
II	Involves two or more lymph nodes in area or on same side of diaphragm
III	Involves lymph node regions on both sides of diaphragm; involves adjacent organ or spleen
IV	Is diffuse disease; least favorable prognosis



Treatment of Hodgkin's Disease

- Both radiation and chemotherapy are used in accordance with the clinical stage of the disease
- Cure is primarily related to the stage of disease at diagnosis
- Long-term prognosis is excellent



Nursing Care of Patients with Hodgkin's Disease

- Mainly directed toward symptomatic relief of the side effects of radiation and chemotherapy
- Education of patient and family
- Malaise is common after radiation therapy, tires easily and child may be irritable and anorexic
- Skin in treated area may be sensitive and must be protected against exposure to sunlight and irritation
- The patient *does not* become radioactive during or after therapy



Emotional Support of the Patient with Hodgkin's Disease

- Support provided should be age-appropriate
- Activity is generally regulated by the patient
- Appearance of secondary sexual characteristics and menstruation may be delayed in pubescent patients
- Sterility is often a side effect of treatment



Nursing Care of the Chronically Ill Child



Chronic Illness

- Behavior problems are lessened when patients can verbalize specific concerns with persons sensitive to their problems
- If they feel rejected by and different from their peers, they may be prone to depression
- Nurses must develop an awareness of the adolescent's particular fears of forced dependence, body invasion, mutilation, rejection, and loss of face, especially within peer groups
- Important to recognize the adolescent's need for self-determination



Developmental Disabilities

- Children with developmental disabilities may often be overprotected, unable to break away from supervision, and deprived of necessary peer relationships
- The pubertal process with its emerging sexuality concerns parents and may precipitate a family crisis



Home Care

- Home health care and other community agencies work together to provide holistic care
- Respite care is sometimes provided to relieve parents of the responsibility of caring for the child



Providing Home Health Care

- Observe how the parents interact with the child
- Do not wait for the child to cry out for attention
- Watch for facial expression and body language
- Post signs above the bed denoting special considerations, such as “never position on left side”
- Listen to the parents and observe how they attend to the physical needs of the child
- Don't be afraid to ask questions or discuss apprehensions
- Be attuned to the needs of other children in the home



Care of the Chronically Ill Child

- Focusing on what the child can do and providing successful experiences are more effective than focusing on the disability
- Involvement of the entire family with the care of the chronically ill child aids in normal family interaction
- Child should be integrated into rather than isolated from the community and society
- The wellness of the child should be the center of the child's life, rather than the disability



Nursing Care of the Dying Child



Facing Death

- The nurse must understand
 - The grieving process
 - Personal and cultural views concerning that process
 - The views of a parent losing a child
 - Perceptions of the child facing death



Facing Death (*cont.*)

- The response to a child's death is influenced by whether there was a long period of uncertainty before the death or whether it was a sudden unexpected event



Facing Death (*cont.*)

- The nurse must
 - Show compassion
 - Demonstrate a nonjudgmental approach
 - Be sensitive and effective in the provision of care
 - Facilitate the grief process by anticipating psychological and somatic responses while maintaining open lines of communication
 - Support the family's efforts to cope, adapt, and grieve
 - Know that hostility is a normal response and may drive away those who do not understand its normalcy in the acute grieving process



Self-Exploration

- How nurses have or have not dealt with their own losses affects present lives and the ability to relate to patients
- Nurses must recognize that *coping is an active and ongoing process*
- An active support system consisting of nonjudgmental people who are not threatened by natural expression of feeling is crucial



The Child's Reaction to Death

- Cognitive development, rather than chronological age, affects the response to death
- Children younger than 5 years of age are mainly concerned with separation from their parents and abandonment
- Preschool children respond to questions about death by relying on their experience and by turning to fantasy
- Children do not develop a realistic concept of death as a permanent biological process until 9 or 10 years of age



The Child's Awareness of His or Her Condition

- Failure to be honest with children leaves them to suffer alone, unable to express their fears and sadness or even to say goodbye



Physical Changes of Impending Death

- Cool, mottled, cyanotic skin and the slowing of all body processes
- Loss of consciousness, but hearing may still be intact
- Rales in the chest may be heard, which are caused by increased pooling of secretions in the lungs
- Movement and neurological signs lessen



Stages of Dying and the Nurse's Role

- Stages
 - Denial
 - Anger
 - Bargaining
 - Depression
 - Acceptance
- Nurse's Role
 - Listen
 - Provide privacy
 - Provide therapeutic intervention
 - Provide information
 - Use appropriate phrases and open-ended statements



Question for Review

- Why are platelets usually not given in patients with idiopathic thrombocytopenic purpura (ITP)?



Review

- Objectives
- Key Terms
- Key Points
- Online Resources
- Review Questions