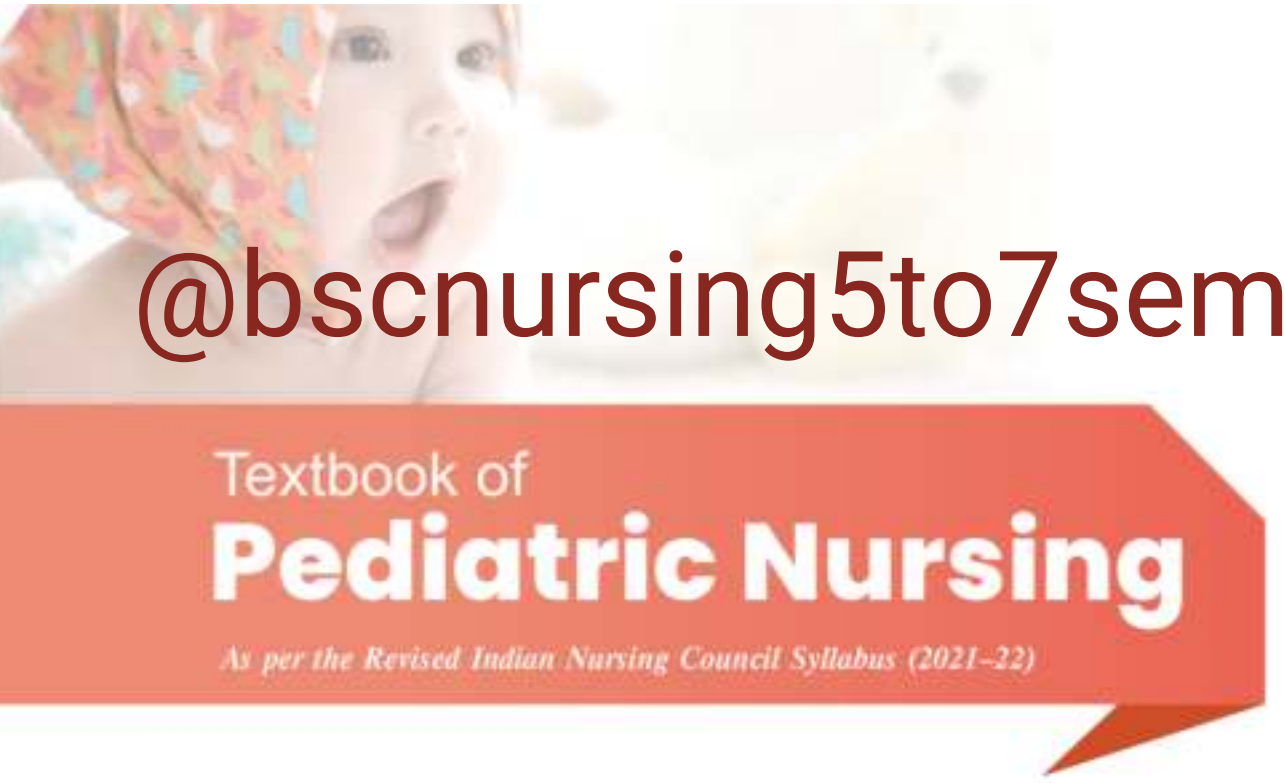

Textbook of Pediatric Nursing

As per the Revised Indian Nursing Council Syllabus (2021–22)

Chapter 16

Hematological Disorders

Chapter Outline

- Introduction
- Blood
- Classification of Blood Disorders

INTRODUCTION

Pediatric blood disorders involve problems with RBCs, WBCs, platelets, blood vessels, bone marrow, lymph nodes, or the proteins involved in bleeding and clotting. These blood disorders affect the functioning and quality of life of infants and children.

BLOOD

Blood contains plasma. Plasma is the liquid component of the blood in which the following blood cells are suspended:

- Red blood cells (erythrocytes) which carry oxygen from the lungs to the rest of the body.
- White blood cells (leukocytes) which help fight infections and aid in the immune process. Types of white blood cells include Lymphocytes, Monocytes, Eosinophils, Basophils, and Neutrophils.
- Platelets (thrombocytes) which help in blood clotting.

Hematopoiesis

Blood cells are made in the bone marrow which is the spongy material in the center of the bones. Other organs and systems that help regulate blood cells are lymph nodes, spleen, and liver which help regulate the production, destruction, and differentiation of cells. The production and development of new cells in the bone marrow is a process called **hematopoiesis**. Blood cells are formed in the bone marrow as a stem cell. A *stem cell* (or hematopoietic stem cell) is the initial phase of all blood cells which matures into red blood cells, white blood cells, and platelets (Fig. 16.1). Immature blood cells are also called blasts. Some blasts stay in the marrow to mature, and others travel to other parts of the body to develop into mature, functioning blood cells.

CLASSIFICATION OF BLOOD DISORDERS

RBC disorders	WBC disorders	Platelets
Anemia Hemolytic anemia, Sickle cell anemia, Pernicious anemia, etc.	Leukemia (ALL, AML, CML)	Hemophilia, Thrombocytopenia ITP

Common symptoms of blood disorders are pallor, weakness, and hypoxia. The diagnostic tests done are CBC, LFT, bone marrow biopsy, platelet count, prothrombin time (PT) and partial thromboplastin time (PTT). These disorders can generally be managed by blood transfusions, Iron and folic acid supplementation.

Anemia

Anemia refers to decrease in number of RBCs or less than the normal quantity of hemoglobin in the blood. Because

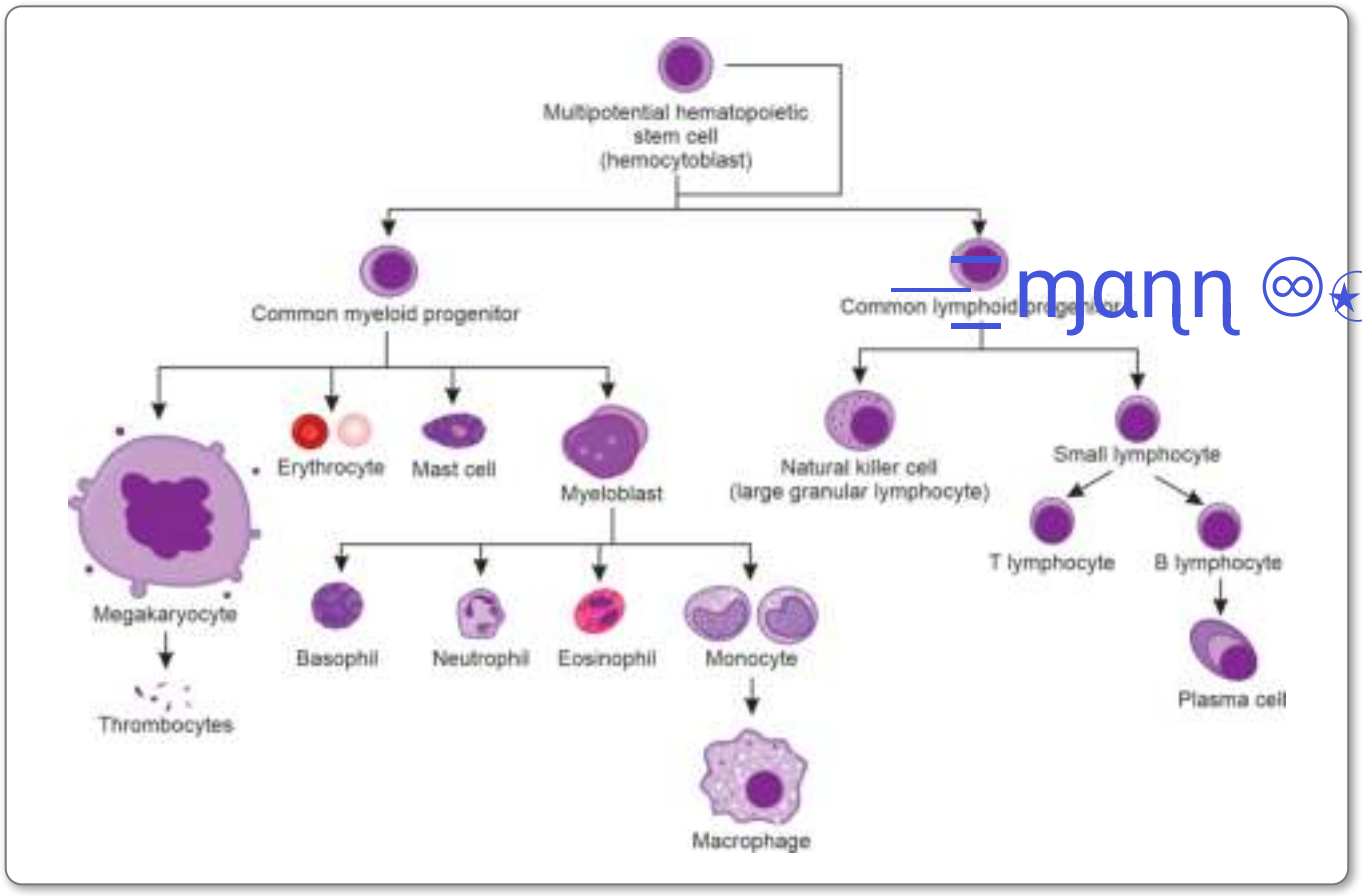

Figure 16.1: The development of different blood cells from hematopoietic stem cell to mature cells

hemoglobin normally carries oxygen from the lungs to the tissues, anemia leads to hypoxia in organs. Since all human cells depend on oxygen for survival, varying degrees of anemia can have a wide range of clinical consequences.

Anemia is the most common disorder of the blood. Anemia can be classified in a variety of ways, based on the morphology of RBCs and underlying etiologic mechanisms. The three main classes of anemia include excessive blood loss (acutely such as a hemorrhage or chronically through low-volume loss), excessive blood cell destruction (hemolysis) or deficient red blood cell production (ineffective hematopoiesis).

Causes

Causes of anemia may be classified as impaired RBC production, increased RBC destruction (hemolytic anemia), blood loss and fluid overload (hypervolemia). Several of these may interplay to cause anemia. The most common cause of anemia is blood loss, but this usually doesn't cause any lasting symptoms unless a relatively impaired RBC production develops, in turn most commonly by iron deficiency.

Signs and Symptoms

Child with anemia report non-specific symptoms of a feeling of weakness, or fatigue, general malaise and sometimes poor concentration. They may also report dyspnea on exertion. In very severe anemia, the body may compensate for the lack of oxygen-carrying capability of the blood by increasing cardiac output. The patient may have symptoms related to this, such as palpitations, angina, intermittent claudication of the legs, and symptoms of heart failure.

- On examination, the signs exhibited may include pallor (pale skin, mucosal linings and nail beds) but this is not a reliable sign. There may be signs of specific causes of anemia, e.g., koilonychia (in iron deficiency), jaundice (in hemolytic anemia), bone deformities (found in thalassemia major) or leg ulcers (seen in sickle-cell disease).
- In severe anemia, there may be signs of a hyperdynamic circulation: Tachycardia, bounding pulse, flow murmurs, and cardiac ventricular hypertrophy. There may be signs of heart failure
- Pica, the consumption of non-edible items such as dirt, paper, wax, grass, ice, and hair, may be a symptom of iron deficiency, although it occurs often in those who have normal levels of hemoglobin
- Chronic anemia may result in behavioral disturbances in children as a direct result of impaired neurological development in infants, and reduced scholastic performance in children of school age.

- Restless legs syndrome is more common in those with iron-deficiency anemia

Diagnosis

Complete blood count is done. Apart from reporting the number of RBCs and the hemoglobin level, the automatic counters also measure the size of the red blood cells by flow cytometry, which is an important tool in distinguishing between the causes of anemia.

The four parameters (RBC count, hemoglobin concentration, MCV and RDW) are measured, allowing others (hematocrit, MCH and MCHC) to be calculated, and compared to values adjusted for age and sex.

Classification of Anemia (Table 16.1)

Red Blood Cell Size

In the morphological approach, anemia is classified by the size of red blood cells; this is either done automatically or on microscopic examination of a peripheral blood smear. The size is reflected in the mean corpuscular volume *(MCV)*.

- If the cells are smaller than normal (under 80 fl), the anemia is said to be *microcytic*;
- If they are normal size (80–100 fl), *normocytic*; and
- If they are larger than normal (over 100 fl), the anemia is classified as *macrocytic*.

*Hypochromic means that the red blood cells have less hemoglobin than normal.

Hemolytic Anemia

Hemolysis is the premature destruction of erythrocytes. Breakdown of RBCs produce heme and globin and releases enzyme LDH. Heme is converted into biliverdin, bilirubin, and finally excreted as stercobilin in feces and urobilinogen in urine. Due to increased hemolysis, there is increase in indirect bilirubin and urobilinogen levels. A hemolytic anemia develops when bone marrow activity cannot compensate for the erythrocyte loss.

Table 16.1: Classification of anemia

Normochromic, normocytic anemia (normal MCHC, normal MCV)	Hypochromic, microcytic anemia (low MCHC, low MCV)	Normochromic, macrocytic anemia (normal MCHC, high MCV)
• Anemias of chronic disease • Hemolytic anemias • Anemia of acute hemorrhage • Aplastic anemias	• Thalassemias • Anemia of chronic disease • Iron deficiency anemia • Lead toxicity associated anemia • Sideroblastic anemia	• Megaloblastic anemia/vitamin B_{12} deficiency • Folate deficiency anemia

Premature erythrocyte destruction can be due to conditions such as intrinsic membrane defects, abnormal hemoglobin, erythrocyte enzymatic defects, and immune destruction of erythrocytes, mechanical injury, and hypersplenism.

Hemolysis may be an extravascular or an intravascular phenomenon. Autoimmune hemolytic anemia and hereditary spherocytosis are examples of extravascular hemolysis because the red blood cells are destroyed in the spleen and other reticuloendothelial tissues.

Causes

Hemolysis can be due to hereditary and acquired disorders. Hereditary disorders include the following:

- Glucose-6-phosphate dehydrogenase (G6PD) deficiency
- Hereditary spherocytosis
- Sickle cell anemia

Acquired causes of hemolysis include the following:

- Immune disorders
- Toxic chemicals and drugs
- Antiviral agents, (e.g., ribavirin)
- Physical damage
- Infections

Signs and Symptoms

Patients with minimal or long-standing hemolytic anemia may be asymptomatic. Clinical manifestations may include the following:

- In intravascular hemolysis, iron deficiency due to chronic hemoglobinuria causes anemia and weakness
- Tachycardia, dyspnea, angina, and weakness occur in patients with severe anemia, as cardiac function is sensitive to anoxia. Angina and heart failure can occur in patients with underlying cardiovascular disease.
- General pallor and pale conjunctivae and fingernails
- Jaundice may occur because of a modest increase in indirect bilirubin in hemolysis.
- Persistent hemolysis may result in the development of bilirubin gallstones; these patients may present with abdominal pain
- Bronze skin color and diabetes occur in hemosiderosis; iron overload may occur in patients who have received multiple transfusions or those who have been administered iron therapy erroneously
- Dark urine may be due to hemoglobinuria
- In addition to hemolysis, patients with thrombotic thrombocytopenic purpura (TTP) may experience fever, neurologic signs, renal failure, and thrombocytopenia.

Diagnosis

Standard blood studies for the workup of suspected hemolytic anemia include the following:

- Complete blood cell count
- Peripheral blood smear

- Serum lactate dehydrogenase (LDH) is elevated
- Serum haptoglobin is decreased as it binds to hemoglobin and removed from body by liver
- Indirect bilirubin is increased.

Management

- Prophylactic folic acid
- Corticosteroids
- Blood transfusion
- Erythropoietin
- Discontinuing medications which cause hemolysis like Penicillin, Ampicillin, Methicillin, Quinine
- Splenectomy removes the major site of red cell destruction and in turn increases the hemoglobin concentration and eliminates the need for an accelerated rate of red cell production. It may be done in case of hereditary spherocytosis.
- Intravenous immunoglobulin G (IVIG) has been used for patients with autoimmune hemolytic anemia.

Sickle Cell Anemia

Sickle cell disease (SCD) is an inherited blood disorder characterized by defective hemoglobin. Hemoglobin consists of four polypeptide chains (two α and two β chains) (Fig. 16.2). Sickle cell anemia occurs due to the replacement of hydrophilic amino acid glutamic acid with hydrophobic amino acid valine at the 6th position of the beta globin chain of hemoglobin. The inclusion of the hydrophobic (non-polar) amino acid promotes non covalent polymerization of hemoglobin resulting in distortion of the RBCs into a sickle shape.

Normal hemoglobin cells are smooth, round, and flexible so they can move through the vessels easily. Sickle cell hemoglobin cells are stiff and sticky, and form into the shape of a sickle, or "C" shape when they lose oxygen (Fig. 16.3). Sickle cells tend to cluster together, and cannot easily move through the blood vessels. It causes a blockage

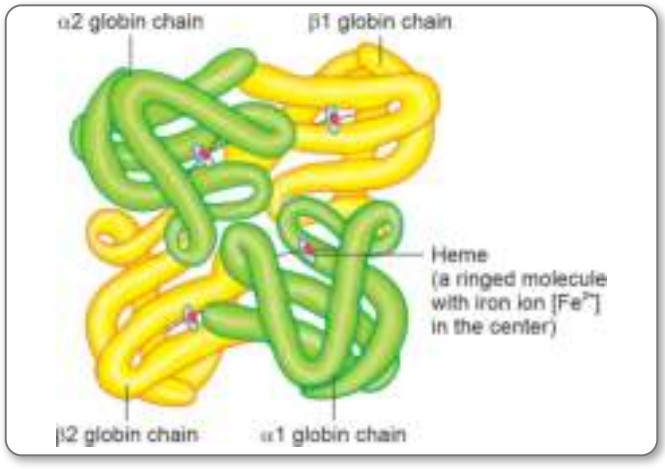

Figure 16.2: Hemoglobin figure

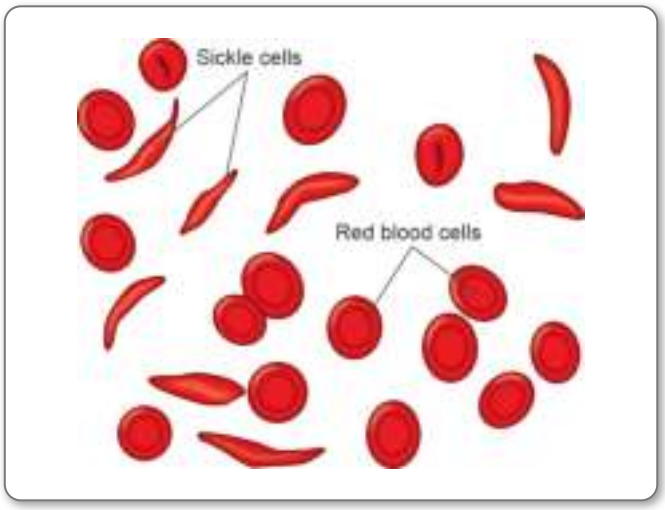

Figure 16.3: Sickled and normal RBCs

in small arteries or capillaries and stops the movement of healthy, normal oxygen-carrying blood. This blockage causes painful and damaging complications of SCD.

A normal hemoglobin can live up to 120 days whereas a sickle cell can live only for about 10–20 days. Also, sickle cells risk being destroyed by the spleen because of their shape and stiffness. Spleen helps filter the blood of infections and sickled cells get stuck in this filter and die. Due to the decreased number of hemoglobin cells circulating in the body, a person with sickle cell disease is chronically anemic. The spleen also suffers damage from the sickled cells blocking healthy oxygen carrying cells and typically infants in the first few years of life. Without a normal functioning spleen, these children are more at risk for infections.

The most common variations of the sickle cell gene are:

- **Sickle cell trait:** It is a carrier state, a heterozygous form characterized by the presence of around 40% HbS and some normal hemoglobin, HbA. This is referred to as HbAS. Children with sickle cell trait usually have no symptoms. Mild anemia may occur and red cells tend to be small. Under intense stressful conditions, exhaustion, hypoxia, or severe infection, the sickling of the defective hemoglobin may occur and result in some complications associated with the sickle cell disease. Most children with the sickle cell trait lead normal lives.
- **Sickle cell anemia:** Most of the normal hemoglobin (HbA) is replaced with the sickle hemoglobin (HbS). This is referred to as HbSS. It is the most common and most severe form of the sickle cell variation. These children suffer from a variety of complications due to the shape and thickness of the sickled cells. Severe and chronic anemia is a common characteristic for children with HbSS.
- **Sickle cell-hemoglobin SC disease:** The child has one copy of both HbS and HbC. This condition is double heterozygote for HbS and HbC characterized by moderate clinical severity.

- **Sickle cell-hemoglobin E disease:** This variation is similar to sickle cell-C disease except that an element has been replaced in the hemoglobin molecule. Some children with hemoglobin E disease are without symptoms. However, under certain conditions, such as exhaustion, hypoxia, severe infection, and/or iron deficiency, some mild to moderate anemia may occur.
- **Hemoglobin S-beta-thalassemia:** This involves an inheritance of both the thalassemia and sickle cell genes. The disorder produces symptoms of moderate anemia and many of the same conditions associated with sickle cell disease. While this disorder more often has milder symptoms than sickle cell disease, it may also produce exacerbations as severe as those of sickle cell disease.

All forms of sickle cell disease can exhibit the complications associated with the disease.

Signs and Symptoms

SCD usually manifests early in childhood. Symptoms include:

- **Acute and chronic pain:** The most common clinical manifestation of SCD is vaso-occlusive crisis causing pain.
- **Bone pain:** Often seen in long bones of extremities primarily due to bone marrow infarction
- Anemia
- **Aplastic crisis:** Serious complication due to infection with B19V
- **Splenic sequestration:** Characterized by the onset of life-threatening anemia with rapid enlargement of the spleen and high reticulocyte count
- **Infection:** With Streptococcus pneumonia
- Growth retardation, delayed sexual maturation, being underweight
- **Hand-foot syndrome (dactylitis):** Presenting as bilateral painful and swollen hands and/or feet in children
- **Acute chest syndrome:** Young children present with chest pain, fever, cough, tachypnea, leukocytosis, and pulmonary infiltrates in the upper lobes.
- **Pulmonary hypertension:** A serious complication of SCD
- **Avascular necrosis of the femoral or humeral head:** This is due to vascular occlusion
- **CNS involvement:** Most severe manifestation is stroke
- **Ophthalmologic involvement:** Ptosis, retinal vascular changes, proliferative retinitis
- **Cardiac involvement:** Dilation of both ventricles and the left atrium
- **GI involvement:** Cholelithiasis is common in children; liver may become involved
- **GU involvement:** Kidneys lose concentrating capacity; priapism is a well-recognized complication of SCD
- **Dermatologic involvement:** Leg ulcers are a chronic painful problem

Triggers of vaso-occlusive crisis include the following:

- **Hypoxemia:** May be due to acute chest syndrome or respiratory complications
- Dehydration
- Changes in body temperature

Diagnosis

There is a typical clinical picture of chronic hemolytic anemia and vaso-occlusive crisis. Electrophoresis confirms the diagnosis with the presence of homozygous HbS and can also document other hemoglobinopathies, (e.g., HbSC, HbS-beta+ thalassemia). Imaging studies like Chest X-ray, Ultrasound, MRI, and CT scan can be done.

The goals of treatment are symptom control and management of disease complications.

Treatment strategies include the following goals:

- **Management of vaso-occlusive crisis:** It is treated with vigorous intravenous hydration and analgesics. Normal saline and 5% dextrose in saline may be used.
- **Management of chronic pain syndromes:** Severe pain can be managed with a strong opioid, (e.g., morphine), and a weak opioid, (e.g., acetaminophen and codeine) for patients with moderate pain. Offer all patients regular acetaminophen and NSAIDs.
- **Management of chronic hemolytic anemia:** Administration of folic acid (0.1–1 mg/day) and RBC transfusion
- **Prevention and treatment of infections:** Neonatal screening, penicillin prophylaxis, appropriate immunizations (particularly against *Streptococcus pneumoniae*), and parental teaching. Recommended parenteral antibiotics include cephalosporins, (e.g., ceftriaxone, cefuroxime) and macrolides for acute chest syndrome. If the patient is discharged, oral antibiotics, (e.g., amoxicillin-clavulanic acid, clarithromycin, cefixime) are useful in selected cases.
- Prevention of stroke
- Detection and treatment of pulmonary hypertension

Nursing Management

- **Impaired gas exchange r/t decreased oxygen carrying capacity of the blood, reduced RBC lifespan/premature destruction as evidenced by dyspnea, confusion, Use of accessory muscles**
  - Monitor respiratory rate, depth, use of accessory muscles, and areas of cyanosis.
  - Auscultate and note the presence or absence of breath sounds and adventitious sounds.
  - Monitor vital signs and note changes in cardiac rhythm.
  - Administer supplemental humidified oxygen as indicated.
  - Perform and assist with chest physiotherapy, intermittent positive-pressure breathing (IPPB) and incentive spirometry.
  - Administer packed RBCs or exchange transfusion as ordered.

- **Ineffective tissue perfusion r/t vaso-occlusive nature of sickling, inflammatory response as evidenced by changes in vital signs: Diminished peripheral pulses/ capillary refill, general pallor**
  - **Carefully monitor vital signs:** Assess pulse points for rate, rhythm, and volume. Take note of hypotension, rapid, weak, and thready pulses, and increased or shallow respirations.
  - Assess skin for pallor, cyanosis, coolness, diaphoresis, and delayed capillary refill.
  - Monitor and note changes in level of consciousness, reports of headache, dizziness, development of sensory and motor deficits (hemiparesis or paralysis), and seizure activity.
  - Maintain adequate fluid intake and monitor urine output.
  - Assess the lower extremities for skin texture, ulcerations, and/or edema as sickling of blood can cause reduced peripheral circulation
  - Maintain room temperature and body warmth
  - Assess lab values, i.e., ABGs, CBC, LDH, AST/ALT, CPK, BUN
  - Administer serum electrolytes. Provide replacements as indicated especially sodium as it is lost during crisis because of fever, diarrhea, vomiting, and diaphoresis. Administer hypo-osmolar solutions (0.45 normal saline) via an infusion pump.
  - Administer hydroxyurea (antisickling agent)
- **Risk for deficient fluid volume r/t hypermetabolic state/ fever, inflammatory processes**
  - Maintain intake and output chart and weigh daily.
  - Note characteristic of urine and specific gravity.
  - Monitor vital signs.
  - Assess patient for fever, changes in level of consciousness, poor skin turgor, dryness of skin and mucous membranes, pain.
  - Closely monitor vital signs during blood transfusions and note presence of dyspnea, crackles, rhonchi, wheezes, jugular vein distention, diminished breath sounds, cough, frothy sputum and cyanosis.
  - Administer IV fluids as indicated.
  - Monitor laboratory studies: Hb/Hct, serum and urine electrolytes.
- **Acute pain r/t intravascular sickling with localized stasis, occlusion and infarction**
  - Assess for pain. Note location, duration, and intensity (scale of 0–10).
  - **Observe nonverbal pain cues:** Gait disturbances, positioning of the body, guarding behavior, facial grimacing, reluctance to move, and physiological manifestations of acute pain (increased BP, tachycardia, increased RR).
  - Teach and discuss alternative pain relief measures: Relaxation techniques, biofeedback, yoga, meditation, progressive relaxation techniques, distraction techniques, guided imagery and breathing techniques.
  - Provide support and carefully position affected extremities.
  - Massage gently affected areas.
  - Encourage ROM exercises.
  - Plan activities during peak analgesic effect.
  - Maintain adequate fluid intake.
  - Apply warm, moist compresses to affected joints and other painful areas. Avoid use of ice or cold compresses.
  - Administer pain medications.
- **Deficient knowledge r/t disease condition as evidenced by questioning**
  - Review precipitating factors, i.e.,
    - Cold environmental temperatures, failure to dress warmly when engaging in winter activities, wearing tight and restrictive clothing which causes peripheral vasoconstriction, which may result in sludging of the circulation, increased sickling, and may precipitate a vaso-occlusive crisis.
    - Stressful situations, strenuous physical activity and contact-type sports, and extremely warm temperatures.
  - Encourage to make more fluids.
  - Encourage ROM exercise and regular physical activity with a balance between rest and activity.
  - Reinforce diet including liver, green leafy vegetables, citrus fruits, and supplementary vitamins such as folic acid.
  - Discuss principles of skin and extremity care and protection from injury. Encourage prompt treatment of cuts, insect bites, sores.
  - Instruct patient to avoid persons with infections such as upper respiratory infections.
  - Inform regarding signs of complications, i.e.,
    - Urine that appears blood tinged or smoky: Sickling in renal medulla
    - Indigestion, persistent vomiting, diarrhea, high fever, excessive thirst: Dehydration leading to vaso-occlusive crisis
    - Severe joint or bone pain: Vaso-occlusive crisis
    - Severe chest pain, with or without cough: Pneumonia, MI
    - Abdominal pain; gastric distress following meals: Cholelithiasis, primarily with bilirubin stones
  - Encourage patient to have routine follow-ups: For CBC, dental examination and ophthalmologic examination.

Aplastic Anemia

Aplastic anemia is a syndrome of bone marrow failure characterized by peripheral pancytopenia and marrow hypoplasia.

Signs and Symptoms

The symptoms are related to the decrease in bone marrow production of hematopoietic cells. The onset is insidious, and the initial symptom is commonly related to anemia or bleeding, although fever or infections may also be noted.

Signs and symptoms of aplastic anemia may include the following:

- Pallor
- Headache
- Palpitations, dyspnea
- Fatigue
- Foot swelling
- Gingival bleeding, petechial rashes (Fig. 16.4)
- Overt and/or recurrent infections
- Oropharyngeal ulcerations

Some of patients with aplastic anemia may present with jaundice and hepatitis.

Diagnosis

Laboratory testing for suspected aplastic anemia includes the following:

- Complete blood count
- Peripheral blood smears
- Hemoglobin electrophoresis and blood-group testing
- Biochemical profile
- Serology for hepatitis and other viral entities
- Autoimmune-disease evaluation for evidence of collagen-vascular disease
- Histocompatibility testing
- Kidney function studies
- Liver function studies
- Transaminase, bilirubin, and lactate dehydrogenase levels
- Bone marrow biopsy

Medical Management

The following medications are used in patients with aplastic anemia:

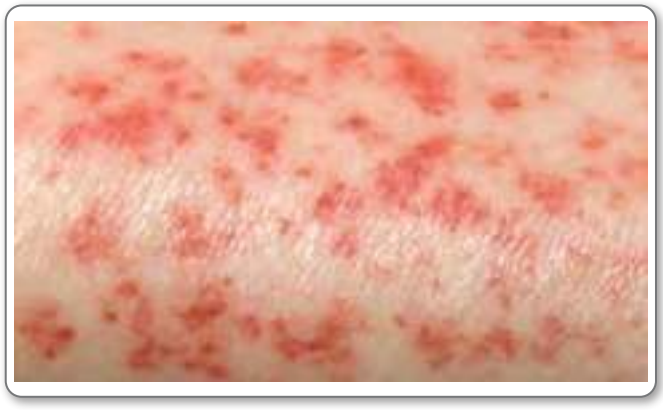

Figure 16.4: Petechial rashes

- Immunosuppressive agents, (e.g., cyclosporine, methylprednisolone, equine anti-thymocyte globulin, rabbit anti-thymocyte globulin, cyclophosphamide, alemtuzumab)
- Hematopoietic growth factors (filgrastim to manage neutropenia)
- Antimetabolite (purine) antineoplastic agents, (e.g., fludarabine)
- Chelating agents, (e.g., deferoxamine which is an iron-chelating agent used to treat iron overload caused by blood transfusions)

Non-pharmacologic management of aplastic anemia includes:

- Supportive care
- Blood transfusions with blood products that have undergone leukocyte reduction and irradiation
- Hematopoietic cell transplantation

Thalassemia

Definition

Thalassemias are inherited disorders of hemoglobin (Hb) synthesis that result from an alteration in the rate of globin chain production. Their clinical severity widely varies, ranging from asymptomatic forms to severe or even fatal entities.

Pathophysiology

Hemoglobin is made of two proteins: Alpha globin and beta globin. Thalassemia occurs when there is a defect in a gene that helps control production of one of these proteins. There are two main types of thalassemia:

- **α-thalassemia:** It occurs when a gene or genes related to the alpha globin protein are missing or changed (mutated).
- **β-thalassemia:** It occurs when similar gene defects affect production of the beta globin protein.

There are many forms of thalassemia. Each type has many different subtypes. Both alpha and beta thalassemia include the following two forms:

1. **Thalassemia major:** Inheritance of defective gene from both parents is essential to develop thalassemia major. Beta thalassemia major is also called **Cooley's anemia**.
2. **Thalassemia minor:** Thalassemia minor occurs if the defective gene is inherited from only one parent. Persons with thalassemia minor are carriers of the disease and usually do not have symptoms.

Symptoms

The most severe form of alpha thalassemia major causes stillbirth. Children born with thalassemia major (Cooley's anemia) are normal at birth, but develop severe anemia during the first year of life. Other symptoms can include:

- Anemia with extreme pallor and enlarged abdomen due to hepatosplenomegaly.

- Bone deformities in the face
- Fatigue
- Growth failure
- Shortness of breath
- Icterus, jaundice

Persons with the minor form of alpha and beta thalassemia have small RBCs (which are identified by looking at their red blood cells under a microscope), but no symptoms.

Diagnosis

- CBC: Low Hb
- Mean corpuscular volume (MCV) and mean corpuscular Hb (MCH) are significantly low
- Hemoglobin electrophoresis shows the presence of an abnormal form of hemoglobin.
- Skeletal survey and other imaging studies reveal classic changes of the bones that are usually encountered in patients who are not regularly transfused.
- ECG and echocardiography are performed to monitor cardiac function.
- HLA typing is performed for patients for whom bone marrow transplantation is considered.
- Eye examinations, hearing tests, renal function tests, and frequent blood counts are required to monitor the effects of deferoxamine therapy and the administration of other chelating agents.

Management

- Treatment for thalassemia major often involves regular blood transfusions and folate supplements. Persons who receive significant numbers of blood transfusions need chelation therapy (Deferoxamine mesylate) to remove excess iron from the body.
- Hematopoietic stem cell transplantation (HSCT)
- Bone marrow transplant
- Splenectomy is the principal surgical procedure used for some patients with thalassemia.

Prognosis

Severe thalassemia can cause early death due to heart failure, usually between ages 20 and 30. Frequent blood transfusions with therapy to remove iron from the body helps improve the outcome. Less severe forms of thalassemia usually have normal lifespan. Genetic counseling and prenatal screening are advised to those with positive family history and who are planning to have children.

Complications

Untreated, thalassemia major leads to heart failure and liver problems, and makes a person more likely to develop infections. Blood transfusions can help control some symptoms, but may result in too much iron which can damage the heart, liver, and endocrine system.

Iron Deficiency Anemia

Iron deficiency anemia is the most common type of anemia. RBCs often appear hypochromic (paler than usual) and microcytic.

Causes

- Insufficient dietary intake or absorption of iron to replace losses from menstruation or losses due to diseases. Iron is an essential part of hemoglobin, and low iron levels result in decreased incorporation of hemoglobin into red blood cells. Anemia may cause poor school performance and lower IQ, abnormal fissuring of the angular sections of the lips (angular stomatitis).
- Iron deficiency anemia can also be due to bleeding lesions of the gastrointestinal tract. Fecal occult blood testing, upper endoscopy and lower endoscopy should be performed to identify bleeding lesions. Worldwide, the most common cause of iron deficiency anemia is parasitic infestation (hookworm, amebiasis, schistosomiasis and whipworm).

Treatment

Treatment for iron deficiency anemia depends on severity and cause.

- Control gastrointestinal bleeding from ulcers or colon cancer.
- Mild to moderate iron-deficiency anemia is treated by oral iron supplementation with ferrous sulfate, ferrous fumarate, or ferrous gluconate. Iron supplements cause stomach upset and/or darkening of the feces. The stomach upset can be alleviated by taking the iron with food; however, this decreases the amount of iron absorbed. Vitamin C aids in the body's ability to absorb iron, so taking oral iron supplements with orange juice is of benefit.
- Deworming: Regime of deworming is as follows:

Age group	Dose and regime
Children 12–59 months of age	Biannual dose of 400 mg albendazole (½ tablet to children 12–24 months and 1 tablet to children 24–59 months)
Children 5–9 years of age	Biannual dose of 400 mg albendazole (1 tablet)
School-going adolescent girls and boys 10–19 years of age Out-of-school adolescent girls 10–19 years of age	Biannual dose of 400 mg albendazole (1 tablet)
Women of reproductive age (non-pregnant, non-lactating) 20–49 years	Biannual dose of 400 mg albendazole (1 tablet)
Pregnant women	One dose of 400 mg albendazole (1 tablet), after the first trimester, preferably during the second trimester

- Vitamin supplements given orally (folic acid) or intramuscularly (vitamin B_{12}) will replace specific deficiencies.
- Recombinant erythropoietin, epoetin alfa, to stimulate red-cell production in anemia of chronic disease, anemia associated with chemotherapy, or anemia associated with renal disease.
- In severe cases of anemia, or with ongoing blood loss, a blood transfusion may be necessary.
- **Hyperbaric oxygen:** Treatment of exceptional blood loss (anemia) is recognized as an indication for hyperbaric oxygen. It is indicated when oxygen delivery to tissue is not sufficient in patients who cannot be transfused for medical or religious reasons, e.g., Jehovah's Witnesses.

Prevention

- Supplement iron rich foods from 4 months of age
- Preterm, LBW- 10–20 mg of elemental iron daily
- Iron supplements during adolescence
- Changing dietary habits
- Wearing shoes to avoid hookworm infection

Megaloblastic Anemia

Megaloblastic anemia, the most common cause of macrocytic anemia, is due to a deficiency of either vitamin B_{12}, folic acid (or both). This deficiency can be due to inadequate intake or insufficient absorption. Folate deficiency normally does not produce neurological symptoms, while B_{12} deficiency does.

- Pernicious anemia is caused by a lack of intrinsic factor. Intrinsic factor is required to absorb vitamin B_{12} from food. A lack of intrinsic factor which may be due to autoimmune condition leads to poor absorption of vitamin B_{12}.
- Macrocytic anemia can also be caused by removal of the functional portion of the stomach, such as during gastric bypass surgery which leads to reduced vitamin B_{12}/folate absorption.
- Other causes of macrocytosis are hypothyroidism, alcoholism, and drugs like Methotrexate, zidovudine that inhibit DNA replication
- In addition to the non-specific symptoms of anemia, specific features of vitamin B_{12} deficiency include peripheral neuropathy and subacute combined degeneration of the cord with resulting balance difficulties from posterior column spinal cord pathology. Other features may include a smooth, red tongue and glossitis.

Diagnosis

The Schilling test, a blood test is done to evaluate the body's ability to absorb vitamin B_{12}.

Management

Diet: Encourage diet high in vitamin B_{12} and folate. Vitamin B_{12} is a nutrient found in some foods like meat, fish, eggs, and milk. Folate is found in foods like beef liver, spinach, and Brussels sprouts.

Nursing Management of Children with Anemia

- **Fatigue r/t decreased hemoglobin and diminished oxygen-carrying capacity of the blood as evidenced by inability to maintain usual level of physical activity.**
  - Assess the specific cause of fatigue
  - Assess the client's ability to perform activities of daily living, and the demands of daily living.
  - Monitor hemoglobin, hematocrit, RBC counts, and reticulocyte counts.
  - Inform about energy-conservation techniques like delegating task to others, setting priorities, and clustering activities. Organization and time management can help the client conserve energy and reduce fatigue.
  - Instruct the client about medications that may stimulate RBC production in the bone marrow, i.e., recombinant human erythropoietin.
  - Provide supplemental oxygen therapy, as needed. Oxygen saturation should be kept at 90% or greater.
  - Anticipate the need for the transfusion of packed RBCs. Packed RBCs increase oxygen-carrying capacity of the blood.
- **Deficient knowledge r/t unfamiliarity with the disease condition as evidenced by questioning members of health care team.**
  - Assess baseline knowledge of the diagnosis, disease process, possible causative factors, and treatment. Explain the function of blood elements.
  - Explain the importance of the diagnostic procedures like CBC, bone marrow aspiration
  - Instruct client to avoid known risk factors such as alcoholism, exposure to toxic chemicals, dietary deficiencies, and the use of certain medications can affect RBC production.
  - For nutritional deficiency anemia: Explain the importance of vitamin B_{12} replacement. Educate the client and the family regarding food rich in iron, folic acid, and vitamin B_{12}. Clients need to have an adequate intake of dark-green leafy vegetables, animal products, including fish, meat, poultry, eggs, milk, and fortified breakfast cereals.
  - **For aplastic anemia:** Explain about blood transfusions, rapid human leukocyte antigen (HLA) typing, immunosuppressive therapy and stem cell transplantation.
- **Risk for infection r/t bone marrow malfunction.**
  - Assess for local or systemic signs of infection, such as fever, chills, swelling, pain, and body malaise.
  - Monitor WBC count.
  - Anticipate the need for antibiotic, antiviral, and antifungal therapy.
  - Instruct the client to avoid contact with people with existing infections.

- If the absolute neutrophil count is less than 500/mm^3, place the client in protective isolation in a private room, limiting visitors, and having all people who come in contact with the client use mask, gown, and gloves.
- Instruct the client to avoid eating raw fruits and vegetables and uncooked meat as these food items can harbor bacteria.
- Stress the importance of daily hygiene, mouth care, and perineal care.
- Teach the client and visitors the proper hand washing.
- Administer WBC growth factor to stimulate the production of neutrophils, i.e., Colony-stimulating factors (CSFs), long-acting pegfilgrastim, filgrastim

- **Risk for bleeding r/t bone marrow malfunction.**
  - Assess the skin for bruises and petechiae. Bruises and petechiae is usually evident when the platelet count drops to 20,000 mm^3.
  - Assess for any frank bleeding from the nose, gums, vagina, or urinary or gastrointestinal tract.
  - Monitor platelet count.
  - Monitor stool and urine for occult blood.
  - Combine laboratory blood sampling test as frequent sampling can cause anemia
  - Encourage diet high in fiber and drinking a lot of fluids to avoid constipation or using a stool softener and other laxatives.
  - Instruct the client about bleeding precautions: Use an electric shaver, not a razor. Use a soft toothbrush when brushing the teeth. Avoid rectal procedures such as suppositories, enemas, and rectal temperature readings.
  - Anticipate the need for a platelet transfusion once the platelet count drops to a very low value to reduce the risk of bleeding. Premedication with antihistamine and antipyretics should be done to reduce transfusion reaction side effects.
- **Activity Intolerance R/T** imbalance between oxygen supply and demand as evidenced by generalized weakness
  - Maintain oxygen saturation.
  - Assist in ADLs
  - Administer blood components (commonly packed RBCs) via intravenous catheter as prescribed.

Hemophilia

Hemophilia refers to a group of bleeding disorders in which it takes a long time for the blood to clot. Hemophilia is the name of any of several hereditary genetic illnesses that impair the body's ability to control bleeding. **Hemophilia A** is a recessive X-linked genetic disorder involving a lack of functional clotting Factor VIII and represents 80% of hemophilia cases (Fig. 16.5).

Hemophilia B is a recessive X-linked genetic disorder involving a lack of functional clotting Factor IX. It comprises approximately 20% of hemophilia cases and **Hemophilia C** is an autosomal genetic disorder.

Causes

Hemophilia is caused by a lack of factor VIII or IX. In most cases, hemophilia is passed down through families (inherited). It most often affects males and females are carriers.

Pathophysiology

When there is bleeding, the body starts a series of reactions that help the blood clot. This is called the coagulation cascade.

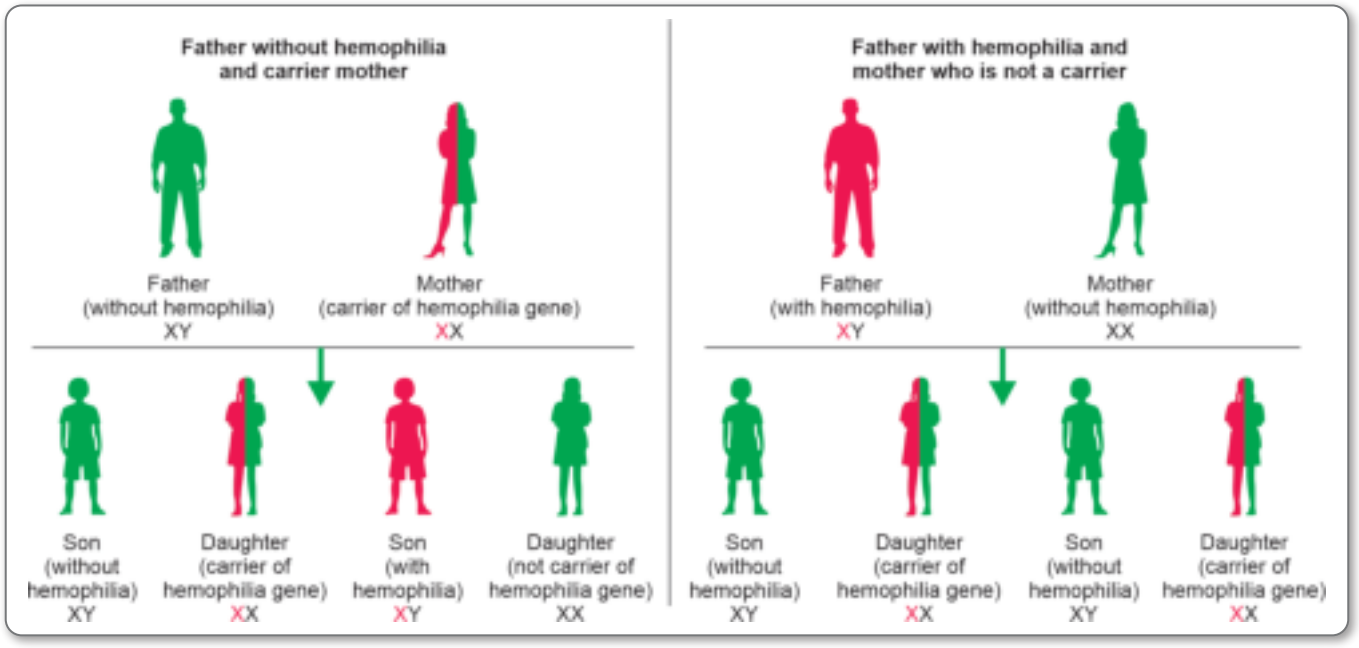

Figure 16.5: Hemophilia inheritance

The process involves 12 coagulation factors. When one or more of these clotting factors are missing, there is a higher chance of bleeding.

Thus when a blood vessel is injured, a temporary scab forms, but the missing coagulation factors prevent fibrin formation, which is necessary to maintain the blood clot. A hemophiliac can bleed for a much longer time. In severe hemophiliacs, even a minor injury can result in blood loss lasting days or weeks, or even never healing completely. In areas such as the brain or inside joints, this can be fatal or permanently debilitating. The bleeding can be external, if the skin is broken by a scrape, cut or abrasion, or it can be internal, into muscles, joints or hollow organs. It might, therefore, present visibly as skin bruises, or subtly as melena hematuria, or bleeding in the brain.

Forms

- **Hemophilia A:** Factor VIII deficiency also known as "Classic hemophilia" (X-linked recessive)
- **Hemophilia B:** Factor IX deficiency also called "Christmas disease" (X-linked recessive)
- **Hemophilia C:** Factor XI deficiency (Ashkenazi Jews, autosomal recessive) sometimes known as "the royal disease".

Symptoms

Children with mild to moderate hemophilia may not have any signs or symptoms at birth especially if they do not undergo circumcision. First symptoms noted are frequent and large bruises and hematomas from frequent bumps and falls as child learns to walk. Swelling and bruising from bleeding in the joints, soft tissue, and muscles may also occur. Children with mild hemophilia may not have noticeable symptoms for many years. Often, the first sign in very mild hemophiliacs is heavy bleeding from a dental procedure, an accident, or surgery. Females who are carriers usually have enough clotting factors from their one normal gene to prevent serious bleeding problems, though some may present as mild hemophiliacs.

Complications

Complications are usually seen in severe and moderate hemophiliacs. Complications may be either directly from the disease or from its treatment:

- Deep internal bleeding, e.g., deep-muscle bleeding, leading to swelling, numbness or pain of a limb.
- Joint damage from hemarthrosis (hemophilic arthropathy), potentially with severe pain, disfigurement, and even destruction of the joint and development of debilitating arthritis.
- Transfusion transmitted infection from blood transfusions.
- Adverse reactions to clotting factor treatment, including the development of an immune inhibitor which renders factor replacement less effective.
- Intracranial hemorrhage, a serious medical emergency caused by the buildup of pressure inside the skull. It can cause disorientation, nausea, loss of consciousness, brain damage, and death.

Management

- Standard treatment involves replacing the missing clotting factor through a vein
- Though there is no cure for hemophilia, it can be controlled with regular injections of the deficient clotting factor, i.e., factor VIII in hemophilia A or factor IX in hemophilia B.
- Some hemophiliacs develop antibodies (inhibitors) against the replacement factors given to them, so the amount of the factor has to be increased or non-human replacement products must be given, such as porcine factor VIII.

Nursing Management

- **Control bleeding:**
  - Assess for internal bleeding.
  - Provide bed rest during bleeding episodes
  - Avoid analgesics like aspirin, IM injections.
  - Administer clotting factors prior to invasive procedures.
  - Assess PTT (PTT is prolonged because of a deficiency in the clotting system factors. The prothrombin time will be normal).
  - Administer DDAVP, it is an analogue of vasopressin and is available intravenously and intranasally.
  - Administer tranexamic acid or epsilon aminocaproic acid as indicated (second-line antifibrinolytic drugs that help hold clots in place by stopping plasmin activity).
  - Anticipate the need for blood replacements. Keep volume expanders and O-negative blood ready.
- **Prevent injury:** Keep side rails padded, provide soft toys. Teach to avoid contact sports; engage in activities such as golf, swimming.
- **Prevent joint damage:** RICE the affected joints: Rest, ice, compression, elevation (hemarthrosis). Maintain normal weight. Encourage for regular exercise and physical therapy.
- Assist with coping with chronic disease
- Recommend genetic testing and counseling for families with hemophilia. Prenatal testing, such as amniocentesis, is available to pregnant women who may be carriers of the condition.

Thrombocytopenia

A normal human platelet count ranges from 150,000 to 450,000 platelets per microliter of blood. One common definition of thrombocytopenia is a platelet count below 50,000 per microliter. Thrombocytopenia is any disorder in which there is an abnormally low amount of platelets.

Causes

Thrombocytopenia is often divided into three major causes of low platelets:

1. Low production of platelets in the bone marrow
2. Increased breakdown of platelets in the bloodstream (called intravascular)
3. Increased breakdown of platelets in the spleen or liver (called extravascular)

Disorders that involve low production in the bone marrow like Aplastic anemia, Cancer in the bone marrow, Cirrhosis of liver, Folate deficiency , etc. Use of certain drugs may also lead to a low production of platelets in the bone marrow. The most common example is chemotherapy treatment. Disorders that involve the breakdown of platelets like DIC and ITP.

Symptoms

- Bruising
- Nosebleeds or bleeding in the mouth and gums
- Rash (pinpoint red spots called petechiae)

Diagnosis

Laboratory tests might include full blood count, liver enzymes, renal function, vitamin B_{12} levels, folic acid levels, erythrocyte sedimentation rate, peripheral blood smear and blood clotting studies (PTT and PT). Other tests that may help diagnose this condition may include Bone marrow aspiration or biopsy and Platelet associated antibodies.

Management

- Treatment depends on the cause of the condition like discontinuing suspected drugs that cause thrombocytopenia, or treating underlying sepsis.
- In some cases, a transfusion of platelets may be required to stop or prevent bleeding.
- Corticosteroids may be used to increase platelet production.
- Lithium carbonate or folate may also be used to stimulate the bone marrow production of platelets

Idiopathic Thrombocytopenic Purpura (ITP)

ITP is the condition of having an abnormally low platelet count (thrombocytopenia) of unknown cause (idiopathic). Most incidents of ITP appear to be related to the production of antibodies against platelets, often ITP is asymptomatic and can be discovered incidentally, but a very low platelet count can lead to an increased risk of bleeding and purpura.

Causes/Pathophysiology

In many cases, cause is not idiopathic but autoimmune, with antibodies against platelets being detected in approximately 60 percent of patients. The stimulus for auto-antibody production in ITP is probably abnormal T cell activity.

Sign and Symptoms

Visible symptoms of ITP include the spontaneous formation of bruises (purpura) and petechiae, especially on the extremities, bleeding from the nostrils, bleeding at the gums, and menorrhagia, any of which may occur if the platelet count is below 20,000 per μL. A very low count (<10,000 per μL) may result in the spontaneous formation of hematomas in the mouth or on other mucous membranes. Bleeding time from minor lacerations or abrasions is usually prolonged.

Serious complications due to an extremely low count (<5,000 per μL) may include subarachnoid or intracerebral hemorrhage, lower gastrointestinal bleeding or other internal bleeding. An ITP patient with an extremely low count is vulnerable to internal bleeding caused by blunt abdominal trauma, as might be experienced in a motor vehicle crash. These complications are not likely in a patient whose platelet count is above 20,000 per μL.

Diagnosis

An ITP is diagnosed with a complete blood count. In some situations, additional investigations (such as a bone marrow biopsy) may be necessary to ensure that the platelet count is not decreased due to other reasons.

Medical Management

Steroids: The first line of treatment usually consists of steroids that suppress the immune system. The dose and mode of administration of steroids is determined by the platelet count and whether there is active bleeding. In emergencies, infusions of dexamethasone or methylprednisolone may be used, while in milder forms the treatment may consist of oral prednisolone. Once the platelet count has improved, the dose of steroid is gradually reduced while monitoring for relapses.

Anti-D: Another strategy that is suitable for Rh-positive patients is treatment with Rho (D) immune globulin (Anti-D). It is intravenously administered which is normally administered to Rh-negative women during pregnancy and after the birth of an Rh-positive infant to prevent sensitization to the Rh factor in the newborn.

Steroid-sparing agents: Immunosuppresants such as mycophenolate mofetil and azathioprine. Intravenous immunoglobulin (IVIg) may be infused in some cases. IVIg can increase the count and reduce bleeding risk.

Thrombopoietin receptor agonists: Thrombopoietin receptor agonists are pharmaceutical agents that treat ITP by stimulating platelet production instead of attempting to curtail platelet destruction, e.g., Romiplostim and Eltrombopag.

Surgical Management

Splenectomy may be considered, as platelets targeted for destruction will usually end up in spleen. The procedure is potentially risky in ITP cases due to the increased possibility of significant bleeding during surgery.

Leukemia

Leukemia is a type of cancer of the blood or bone marrow characterized by an abnormal increase of immature white blood cells called 'blasts'. Leukemia is a broad term covering a spectrum of diseases affecting the blood, bone marrow, and lymphoid system, which are all known as hematological neoplasms.

Causes

Leukemia results from mutations in the DNA. Certain mutations can trigger leukemia by activating oncogenes or deactivating tumor suppressor genes, and thus disrupting the regulation of cell death, differentiation or division. These mutations may occur spontaneously or as a result of exposure to radiation or carcinogenic substances.

Pathophysiology

In developing embryos, blood formation occurs in the yolk sac. As development progresses, blood formation occurs in the spleen, liver and lymph nodes. When bone marrow develops, it eventually assumes the task of forming most of the blood cells for the entire organism.

Hematopoietic stem cells (HSCs) reside in the bone marrow and have the ability to give rise to all of the different mature blood cell types and tissues as explained in Figure 16.1 earlier.

All blood cells are divided into three lineages.

1. Red blood cells also called erythrocytes which are the oxygen-carrying cells.
2. Lymphocytes are the keystone of the adaptive immune system. They are derived from common lymphoid progenitors. The lymphoid lineage is composed of T-cells, B-cells and natural killer cells
3. Cells of the myeloid lineage include granulocytes, megakaryocytes and macrophages.

Types

The leukemias are categorized by subtype into two major classifications: acute lymphoid leukemia (ALL)/blast stem leukemia, the most common type in children, and acute myelogenous leukemia (AML), most frequent in adults and the most common form of leukemia overall. The primary difference is the type of WBC involved. In ALL, the immature WBCs are lymphocytes, and in AML, the WBCs involved are cells from the myeloid line, primarily granulocytes or monocytes.

Four major kinds of leukemia		
Cell type	**Acute**	**Chronic**
Lymphocytic leukemia (or "lymphoblastic")	Acute lymphoblastic leukemia (ALL)	Chronic lymphocytic leukemia (CLL)
Myelogenous	Acute myelogenous leukemia (AML) (or Myeloblastic)	Chronic myelogenous leukemia (CML)

- **Acute leukemia** is characterized by a rapid increase in the number of immature blood cells. Crowding due to such cells makes the bone marrow unable to produce healthy blood cells. Immediate treatment is required in acute leukemia due to the rapid progression and accumulation of the malignant cells, which then spill over into the bloodstream and spread to other organs of the body. Acute forms of leukemia are the most common forms of leukemia in children.
- **Chronic leukemia** is characterized by the excessive buildup of relatively mature, but still abnormal, white blood cells. It takes months or years to progress, the cells are produced at a much higher rate than normal, resulting in many abnormal white blood cells. Whereas acute leukemia must be treated immediately, chronic forms are monitored for some time before treatment to ensure maximum effectiveness of therapy. Chronic leukemia mostly occurs in older people, but can theoretically occur in any age group.

Acute Lymphoblastic Leukemia

Acute lymphoblastic leukemia (**ALL**) is a form of cancer of the white blood cells characterized by excess lymphoblasts. Malignant, immature WBCs continuously multiply and are overproduced in the bone marrow. ALL causes damage and death by crowding out normal cells in the bone marrow, and by infiltrating to other organs.

Therefore, people with ALL experience symptoms from malfunctioning of the erythrocytes, leukocytes, and platelets. Impact of leukemia on other blood cells is shown in Table 16.2.

Table 16.2: Impact of leukemia on blood cells

Cell type	Compromise	Resulting impact	Residual effects
Red blood cells (RBCs)	Low production (anemia)	Decreased oxygen transport	Shortness of breath with exertion Active intolerance
			Pallor
Platelets	Decreased production	Decreased clotting	Bleeding tendency
White blood cells (WBCs)	Decreased functional WBCs	Decreased immune function	Susceptibility to infection
Immature leukocytes	Excessive production of nonfunctional cells	Crowding within bone and pressure	Pain in bone and joints
		Thinning of bone shaft and weakening	Pathologic fractures

The signs and symptoms of ALL are:

- Generalized weakness and fatigue
- Anemia
- Frequent or unexplained fever and infection
- Weight loss and/or loss of appetite
- Excessive and unexplained bruising
- Bone pain and joint pain are caused by the spread of "blast" cells to the surface of the bone or into the joint from the marrow cavity
- Breathlessness
- Enlarged lymph nodes, liver and/or spleen
- Pitting edema in the lower limbs and/or abdomen
- Petechiae are tiny red spots or lines in the skin due to low platelet levels

Diagnosis

- Medical history, physical examination, complete blood count, and blood smears to see blast cells.
- A bone marrow biopsy is conclusive proof of all.
- A lumbar puncture to know if CNS is involved.
- Pathological examination, cytogenetics (in particular the presence of Philadelphia chromosome) and immunophenotyping, establish whether the myeloblastic (neutrophils, eosinophils, or basophils) or lymphoblastic (B lymphocytes or T lymphocytes) cells are the problem.
- Ultrasound or CT scanning to find invasion of other organs commonly the lung, liver, spleen, lymph nodes, brain, kidneys, and reproductive organs.

Treatment

Management of ALL focuses on control of bone marrow and systemic disease. Additionally, treatment must prevent leukemic cells from spreading to other sites, particularly the CNS by performing lumbar punctures. In general, ALL treatment is divided into several phases.

Chemotherapy

- *Induction chemotherapy* to bring about bone marrow remission. For children with low-risk ALL, standard therapy usually consists of three drugs (prednisone, L-asparaginase, and vincristine) for the first month of treatment.
- *Consolidation therapy* or *intensification therapy* to eliminate any remaining leukemia cells. There are many different approaches to consolidation, but it is typically a high-dose, multi-drug treatment that is undertaken for a few months. Patients with low- to average-risk ALL receive therapy with antimetabolite drugs such as methotrexate and 6-mercaptopurine (6-MP). High-risk patients receive higher drug doses of these drugs, plus additional drugs.
- *CNS prophylaxis* (preventive therapy) to stop the cancer from spreading to the brain and nervous system in high-risk patients. Standard prophylaxis may include radiation of the head and/or drugs delivered directly into the spine.
- *Maintenance treatments* with chemotherapeutic drugs to prevent disease recurrence once remission has been achieved. Maintenance therapy usually involves lower drug doses, and may continue for up to three years.

Bone marrow transplantation is used to treat children with ALL and AML. The goal of BMT is to replace a child's diseased bone marrow with healthy bone marrow. It is reserved for second remission with ALL. It may be used with first AML remission to improve prognosis.

There are three types of BMT based on how the healthy stem cells are collected.

1. **Autologous bone marrow transplant:** Stem cells can be collected in child in two ways:
   i. **Peripheral blood stem cells (PBSCs):** Stem cells are taken from child by apheresis. Apheresis is a process of collecting stem cells that float in the blood (peripheral blood stem cells). The cells are then given back to child after intensive treatment.
   ii. **Bone marrow harvest:** Stem cells are collected from child by a needle placed into the soft center of the bone. Most sites used for bone marrow harvesting are in the hip bones because they have a larger number of stem cells.
2. **Allogeneic bone marrow transplant:** The donor is another person who shares the same or similar genetic type as the child. This is often a brother or sister. Finding a matching donor can be a lengthy process. In some cases, a parent may be a donor. Or the donor may be a matched unrelated donor found on a bone marrow registry. Stem cells are taken from the donor either by apheresis or bone marrow harvest.
3. **Umbilical cord blood transplant:** Stem cells are taken from an umbilical cord right after a baby's birth. These stem cells grow into mature blood cells quicker and more effectively than stem cells from the bone marrow of another child or adult. The stem cells are tested, typed, counted, and frozen until they are needed for a transplant.

Before BMT, through physical exams and tests it is ensured that functions heart, kidney, liver, and lungs are normal. Post BMT, provide supportive care to prevent and treat infections, side effects, and physical and emotional complications.

Radiation Therapy

Radiation therapy is used on painful bony areas, in high disease burdens, or as part of the preparations for a bone marrow transplant (total body irradiation). Radiation in the form of whole-brain radiation is also used for CNS prophylaxis, to prevent recurrence of leukemia in the brain. Whole-brain prophylaxis radiation used to be a common method in treatment of children's ALL. Recent studies show that CNS chemotherapy is more favorable and has less developmental side-effects. As a result, the use of whole-brain radiation has been more limited.

Nursing Management

- **Risk for infection r/t alterations in mature WBCs**
  - Place in a private room. Limit visitors as indicated. Prohibit live plants or flowers. Restrict fresh fruits and make sure they are properly washed or peeled. Coordinate patient care so that leukemic patient doesn't come in contact with staff who also care for patients with infections or infectious diseases.
  - Enforce hand washing protocol for all personnel and visitors.
  - Closely monitor temperature.
  - Encourage frequent turning and deep breathing, to prevent stasis of respiratory secretions, reducing risk of atelectasis or pneumonia.
  - Auscultate breath sounds, noting crackles, rhonchi. Inspect secretions for changes in characteristics: increased sputum production or change in sputum color. Observe urine for signs of infection: Cloudy, foul-smelling, or presence of urgency or burning with voids.
  - Inspect oral mucous membranes. Provide good oral hygiene. Use a soft toothbrush, sponge, or swabs for frequent mouth care.
  - Promote good perianal hygiene. Examine perianal area at least daily during acute illness. Provide sitz baths, using Betadine or Hibiclens if indicated. Avoid rectal temperatures, use of suppositories.
  - Administer antibiotics and colony stimulating factors.
- **Risk for deficient fluid volume r/t vomiting, diarrhea, hemorrhage**
  - Monitor I and O. Calculate insensible losses and fluid balance. Note decreased urine output in presence of adequate intake. Measure specific gravity and urine pH.
  - Weigh daily.
  - Explain that chemotherapy may cause weight loss and anorexia so encourage the patient to eat and drink high-calorie, and high-protein foods.
  - Monitor BP and HR. Evaluate skin turgor, capillary refill, and general condition of mucous membranes to assess hypovolemia
  - Administer IV fluids as indicated.
  - Administer medications, like antiemetics (ondansetron), Allopurinol to prevent nephropathy, (Potassium acetate or citrate, sodium bicarbonate to alkalinize urine, Stool softeners).
  - Monitor laboratory studies: Platelets, Hb/Hct, clotting
  - Administer RBCs, platelets, clotting factors.
  - Maintain external central vascular access device (subclavian or tunneled catheter or implanted port).
- **Acute Pain r/t bone marrow packed with leukemic cells**
  - Assess pain.
  - Provide pain relief to address acute pain related to mucositis, bone pain. Administer analgesics as ordered (acetaminophen, opioids).
  - Use non pharmacological interventions.
- **Activity Intolerance r/t generalized weakness**
  - Assess ADLs.
  - Provide quiet environment and uninterrupted rest periods.
  - Recommend small, nutritious, high-protein meals and snacks throughout the day.
  - Provide supplemental oxygen.
- **Deficient knowledge**
  - Review pathology of specific form of leukemia and various treatment options.
  - Provide psychological support by establishing a trusting relationship to promote communication.
  - Explain procedure at child's level of understanding including what will be seen, felt, heard, and smelled; use drawings when appropriate.
  - Reinforce physician's explanation of diagnosis and treatment plan.
- **Body image disturbance**
  - Address potential for disturbed body image related to hair loss due to radiation treatment by helping family plan in advance for hair coverings.
- **Anxiety**
  - Explain treatment protocol to parents and include the child with cancer in planning care, as age permits.
  - Prepare child and family for tests and knowledge of chemotherapy treatment, side effects and complications to monitor.
  - Allow the patient and family to discuss or verbalize their anger and depression.
  - Let the family participate in patient care as much as possible.

Summary

Pediatric blood disorders impact a child's quality of life and, in some cases, can even be life-threatening. These disorders involve the blood and include problems with red blood cells, white blood cells, platelets, bone marrow, lymph nodes, and spleen. Children with blood disorders may have trouble moving oxygen around their body, replacing lost blood, or fighting other diseases. Performing a complete blood count is essential for diagnosis. Treatment includes medications, blood transfusions, and sometimes splenectomy.

Assess Yourself

1. List the common clinical features of anemia.
2. What are the causes of sickle cell crisis?
3. Define thrombocytopenia.
4. Discuss the nursing management of a child with ALL.
5. Megaloblastic anemia is due to deficiency of .....
6. Beta thalassemia major is also called .....
7. Hemophilia is caused by a lack of factor ..... and .....
8. Hemophilia B, factor IX deficiency is also called .....