CHILD WITH BLOOD DYSFUNCTION

Classifications of Hematology Disorders

A-Inherited disorders

Thalassemia –sickle cell anemia and hemophilia

B-Acquired disorders

Iron deficiency anemia – Megaloplastic anemia

C-Neoplasm (Malignancy)

Leukemia –plastic anemia

Anemia: is a reduction in RBCs mass and/or hemoglobin concentration compared with normal values for age.

Causes of Anemia

1. Nutritional deficiency – iron, foliate, B12.
2. Increased destruction of RBCs – sickle cell anemia.
3. Impaired or decreased rate of production – plastic anemia.
4. Excessive blood loss – hemophilia.

Classification of anemia:

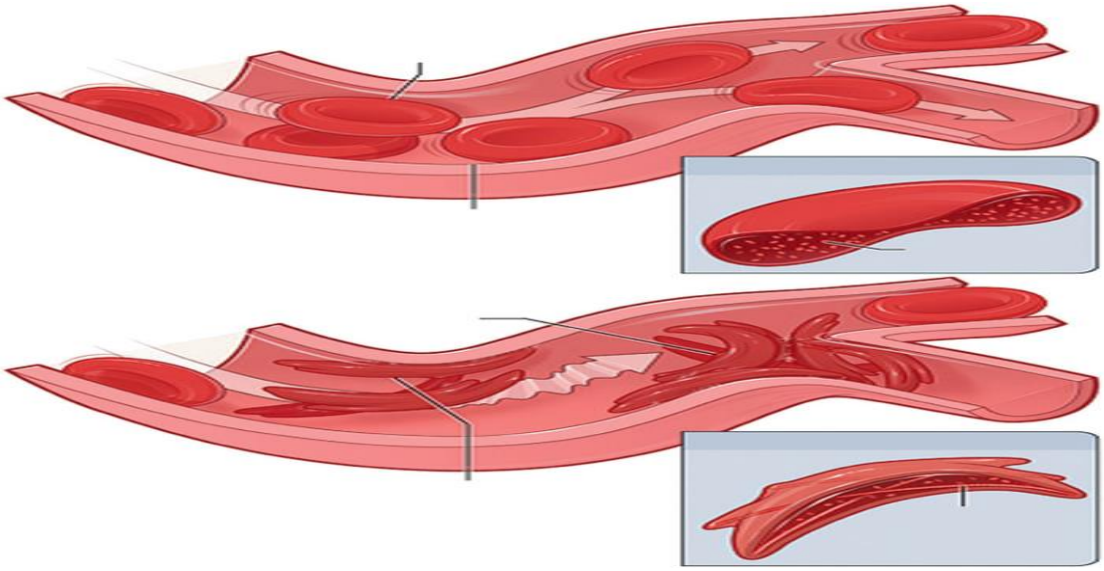
1. Decreased Production of RBCs:
    a. Iron Deficiency Anemia.
    b. A plastic Anemia.
2. Increased Destruction of RBCs:
    a. Hemolytic Anemia.
    b. Sickle Cell Anemia.
    c. Thalassemia.
3. Blood loss:
    a. Iron Deficiency Anemia.
    b. Hemorrhagic Anemia.
4. Morphologic (describe the size of RBCs):
    a. Normocytes (normal cell size).
    b. Microcytes (smaller than normal cell size).
    c. Macrocytes (larger than normal cell size).
5. According to the amount of hemoglobin in the cell (describe the color and Hb content of the cells):
    a. Normochromic (sufficient or normal amount of hemoglobin per RBC).
    b. Hypochromic (reduced amount of hemoglobin per RBC).
    c. Hyperchromic (increased amount of hemoglobin per RBC).
6. According to shape (irregular shape):
    a. Spherocytes (globular cells)
    b. **Drepanocytes (sickle shaped** cells).
    c. Numerous other irregular shaped cells.

Sickle Cell Anemia

Sickle cell disease (SCD) is a hereditary hemoglobinopathy characterized by the partial or complete replacement of normal hemoglobin with abnormal hemoglobin S (**Hgb S**) in red blood cells.

Etiology and Pathophysiology

Sickle cell disease is an autosomal recessive disorder. If both parents have the trait, with each pregnancy the risk of having a child with the disease is 25%.



Clinical manifestation of sickle cell anemia

A. General
   1. Possible growth retardation.
   2. Chronic anemia (Hgb 6 to 9 g/dl).
   3. Possible delayed sexual maturation.
   4. Marked susceptibility to sepsis.

B. Vaso-occlusive (crisis)
   1. Pain in area of involvement.
   2. Manifestations related to ischemia of involved areas:
      a. Extremities (painful swelling of hands and feet (sickle cell dactylitis, or hand-foot syndrome), painful joints).
      b. Abdomen (severe pain resembling acute surgical condition).
      c. Cerebrum (stroke, visual disturbances).
      d. Chest (symptoms resembling pneumonia, protracted episodes of pulmonary disease).
      e. Liver (obstructive jaundice, hepatic coma).
      f. Kidney (hematuria).

C. Sequestration crisis
   1. Pooling of large amounts of blood.
   a. Hepatomegaly.

b. Splenomegaly.
c. Circulatory collapse.

Management and Nursing Care

A -Prevent sickling by:

1. Promote tissue oxygenation and prevent tissue hypoxia through:
2. Promote proper hydration by:
3. Prevent infection by:
4. Control pain by:
   a. Administer analgesic and Comfortable position.
   b. Apply warmth to painful area.
2. Promote rest by keep child at bed rest.
3. Replace blood and observe for signs of transfusion reactions.
4. Prevent psychological problem.
5. Promote long term care and genetic counseling

Hemophilia

Refers to group of bleeding disorders resulting from congenital deficiency of one of the factors necessary for coagulation of the blood.
Most common forms of disorders are factors **VIII classic** or **type A hemophilia**.
Other types are **IX Christmas or type B**. It is more common in males than female.

Modes of transmission

Hemophilia refers to a group of hereditary bleeding disorders that result from a deficiency in specific clotting factors.

1. Hemophilia A, or classic hemophilia, is caused by a deficiency of factor VIII in the blood and occurs in 1 in 5000 male births.
2. Hemophilia B, also known as Christmas disease, is caused by a deficiency of factor IX and occurs in 1 in 25,000– 30,000 male births.

Hemophilia A accounts for 85% of persons with hemophilia, and 10% to 15% have hemophilia B.

Hemophilia A

1. Severe 1% spontaneous bleeding without trauma.
2. Moderate 1% - 5% bleeding with trauma.
3. Mild 5% - 50% bleeding with severe trauma or surgery.

Clinical Manifestations

1. Prolonged bleeding.
2. Epistaxia, bleeding from nose.
3. GIT and spinal cord bleeding, Petechia
4. **Hemoarthrosis: joint bleeding**

Medical management

1. Replacement of the missing clotting factor.
2. **Corticosteroids are given for hematoma**.

Nursing care:

1. Prevent bleeding
2. Recognize and control bleeding (RICE)
   - Rest
   - **Ice**
   - Compression
   - Elevation
3. Client education
4. **NO rectal temperatures**

Thalassemia

Thalassemia is the most common inherited blood disorder characterized by deficiencies in the rate of production of specific **globin chains in hemoglobin**.
The thalassemias are classified according to the hemoglobin chain affected and by the amount of the globin chain that is synthesized.
B-thalassemia is the most common of the thalassemia and occurs in four forms:
two heterozygous forms**,**
thalassemia minor, generally an asymptomatic silent carrier state, and thalassemia trait, which produces mild **Microcytic anemia;**
thalassemia intermedia, which is manifested as Splenomegaly and moderate anemia to severe anemia.
thalassemia major (also known **as Cooley anem**ia) There is a homozygous form, which results in a severe anemia that is not compatible with life without transfusion support.

Clinical manifestation of Thalassemia

- Anemia before diagnosis
  1. Pallor.
  2. **Enlarged spleen/ liver. With progressive anemia**
  3. Unexplained fever.
  4. Poor feeding.

Thalassemia Diagnosis

- Checking a sample of amniotic fluid (which surrounds the baby in the womb) or cells from the placenta
- Clinical feature
- Complete blood count (CBC)
- **Another test may be performed called hemoglobin electrophoresis.**

Therapeutic Management

The advantages of this therapy include:

1. Improved physical and psychologic well-being because of the ability to participate in normal activities.
2. Decreased cardiomegaly and hepatosplenomegaly.
3. Fewer bone changes.
4. Normal or near-normal growth and development until puberty.
5. Fewer infections.

Management of thalassemia

1. Blood Transfusions.
2. Surgery splenectomy.
3. Bone Marrow Transplants.
4. Other Treatments.
5. Antibiotic and vaccine to help prevent infections.
6. Folic acid is a vitamin B9 that helps build red blood cells.

Complications of Thalassemia

1. Growth failure and pubertal delay.
2. Skin change and skeletal abnormalities.
3. Hepatosplenomegaly and Heart failure.
4. Endocrines abnormalities such as (insulin disorder, hypothyroidism).
5. Psychological problems to child and his family such as (daily activity, school problem).

The objectives of nursing care are

1. Promote compliance with transfusion and chelation therapy.
2. Assist the child in coping with the anxiety-provoking treatments and the effects of the illness.
3. Foster the child's and family's adjustment to a chronic illness.
4. Observe for complications of multiple blood transfusions.

Leukemia

Leukemia is a broad term given to a group of malignant diseases of the bone marrow and lymphatic system.

The three main consequences of leukemia are:

1. Anemia from decreased erythrocytes.
2. Infection from neutropenia.
3. Bleeding from decreased platelet production.

Causes of leukemia

1. Radiation.
2. Genetic.
3. Certain kinds of chemicals.
4. The leukemia increase with identical twins.
5. The leukemia associated with disorders of the immune mechanisms of the body.

Signs and Symptoms

- Fever.
- Pallor.
- Overt signs of bleeding.
- Lethargy or malaise.
- Anorexia.
- Large joint or bone pain.
- Petechiae, frank bleeding.
- Enlarged liver or spleen, changes in lymph nodes.
- Neurologic changes.

Leukemia – Diagnosis

- ✓ History & physical.
- ✓ Anemia, thrombocytopenia, and *neutropenia*.
- ✓ Bone Marrow aspiration is the definitive test.

Physical Examination

1. **General**- Orientation, general state of health.
2. **Skin**- Petechiae or ecchymosis, lesions or sores, presence of blood from gum or nose, color of skin
3. **Lungs**- Evidence of infection, dyspnea, Tachypnea.
4. **Abdomen**- Hepato-splenomegaly, mass, decreased bowel sounds.
5. **Neurologic**- Altered consciousness, altered sensation, abnormal reflexes, abnormal cerebellar functions, unstable gait, and cranial nerve deficits.
6. **Lymphatic**- Enlarged lymph nodes.



Treatment

1**. Chemotherapy.**

A. Induction phase Almost immediately after confirmation of the diagnosis, induction therapy is begun and lasts for 4 to 5 weeks.
B. Maintenance therapy, weekly or monthly complete blood counts are taken to evaluate the marrow's response to the drugs.
C. Reinduction After Relapse

2. Bone marrow transplants.

Leukemia – Nursing Management

1. Prepare child/family for diagnostic tests and procedure.
2. Relieve pain.
3. Provide emotional support to child and family.
4. Prevent complications of myelosuppression.
    a. Infection.
    b. Private room.
    c. Strict hand washing.
    d. Restriction of visitors.
    e. Adequate nutrition.
    f. Masks.
    g. Hemorrhage.
    h. Platelet infusions.
    i. Avoid skin puncture.
    j. Mouth care.
5. **No rectal temperatures**
6. Avoid activities that could cause injury.
7. General and frequent nursing assessment for detecting complications of this disease, side effect of chemotherapy and signs and symptoms of infection.
8. When blood transfusion using the nurse explain the purposes and procedure to make child feel better.
9. When child has anemia the nurse keeps him at rest period and balanced meals and give child adequate of protein, CHO and fluid in order to prevent malnutrition and anemia.
10. Antiemetic schedule need to be minimize or prevent the nausea and vomiting.
11. Observe carefully for hemorrhage such as petechia and ecchymosis in the skin.
12. Observe general hygiene such as oral hygiene and skin hygiene for any inflammation.
13. Pain management.
14. Recording of vital signs and body weight.
15. Provide education for child's family about diagnosis, prognosis and treatment of leukemia.
16. Provide continued emotional support for child and his family.