**Hematological Disorders in Children**

Learning objectives

* To know the anatomy and physiology of the hematological system
* Identify differences between child and adult hematological systems.
* To Describe the etiology, pathophysiology, clinical manifestations, diagnosis, and treatment of common hematological alterations
* To Discuss the nursing management and interventions appropriate for children with hematological alterations

**Introduction and Overview of hematological** **System**

**Hematology terminology**

* **Aleukemic Leukemia**: A fatal condition of the blood-forming tissues, characterized by marked proliferation of immature cells in the bone marrow, without their presence, in any great numbers, in the blood steam.
* **Anemia**: A condition in which the blood is deficient in quantity or quality of erythrocytes.
* **Aplastic Anemia**: Anemia characterized b incomplete or effective blood development.
* **Hematocrit**: The packed cell volume (PVC) of red blood cells obtained by globin and forms hemoglobin.
* **Hematology**: The branch of medicine that deals with the study of blood cells, blood producing organs and the manner in which these cells and organs are affected in disease.
* **Hematopoietic (Hemopoietic)**: Blood forming.
* **Hemoglobin**: The coloring matter of the red blood cells. A complex iron-bearing pigment that carries oxygen and carbon dioxide.
* [**leukemia**](http://www.hematology.org/Patients/Cancers/Leukemia.aspx): a type of [cancer](http://www.hematology.org/Patients/Cancers/) found in the blood and bone marrow that is caused by the production of abnormal [white blood cells](http://www.hematology.org/Patients/Blood-Basics/5222.aspx#WhiteBloodCells)
* **Leukocytosis**: An increase in leukocytes in the blood.
* **Leukopenia**: A reduction in the number of leukocytes in the blood.
* **oncology**: the scientific study of cancer
* **red blood cell**: the most common blood cell; it carries the protein hemoglobin, which transports oxygen from the lungs to the rest of the body; red blood cells are also known as erythrocytes or RBC
* **Polycythemia**: An increase in the total number of erythrocytes
* **Purpura**: Small spots on the skin formed by subcutaneous effusion of blood.
* [**sickle cell anemia**](http://www.hematology.org/Patients/Anemia/Sickle-Cell.aspx): a congenital inherited blood disorder characterized by a different type of hemoglobin that causes [red blood cells](http://www.hematology.org/Patients/Basics/#a3) to become rigid and sickle-shaped
* **stem cell:** a cell that has the unique property of self-renewal as well as the ability to develop into other types of specialized cells, such as blood cells
* **Thrombocytopenia**: A decrease in blood platelets; also thrombopenia.
* **bone marrow:** the soft, spongy tissue inside of bones where [blood cells](http://www.hematology.org/Patients/Basics/) are produced
* [**bone marrow transplantation**](http://www.hematology.org/Patients/Basics/): the transfer of healthy bone marrow cells into a person whose bone marrow is defective or has been damaged by chemotherapy or radiation
* [**hemophilia**](http://www.hematology.org/Patients/Bleeding.aspx#a2): a congenital or inherited [bleeding disorder](http://www.hematology.org/Patients/Bleeding.aspx) caused by a shortage of clotting factors in the bloo

**Blood forming.**

* Production of blood cells varies with age
  + By birth, virtually all bone marrow cavities are actively hematopoietic In childhood,hematopoiesis moves to central bones (vertebrae,sternum, ribs, pelvis)

**Components of the Blood**

* Blood cell

-RBCs, WBCs, Platelets

* Plasma : Plasma is about 90% water and 10% solutes ( water, albumin, electrolytes, clotting factors)
* Cellular Components
  + red blood cells *(erythrocytes),*
  + white blood cells *(leukocytes),*
  + platelets *(thrombocytes).*

**RBCs**

* + carry hemoglobin which is attached to oxygen- provides O2 to the tissues
  + life span 120 days
  + manufacture regulated by erythropoietin

**Differences between child and adult hematological systems**

* In infants and young children all of the bone contains red marrow (so-called because of its color from formation of erythrocytes).
* At the end of adolescence, only the ribs, sternum, vertebrae, and pelvis continue to produce blood cells. The remainder of the bone marrow becomes yellow from deposition of fat.

**Classifications of Hematology Disorders**

**A-Inherited disorders**

Thalassemia –sickle cell anemia and hemophilia

**B-Acquired disorders**

Iron deficiency anemia –megaloplasticanemia

**C-Neoplasm (Malignancy)**

Leukemia –aplastic anemia

**Anemia**: A condition in which the blood is deficient in quantity or quality of erythrocytes(RBC).

**Causes of Anemia**

* Nutritional deficiency – iron, folate, B12
* Increased destruction of RBCs – sickle cell anemia
* Impaired or decreased rate of production – aplastic anemia
* Excessive blood loss – hemophilia
* The basic cause of anemia is either

An etiologic classification is based on the various conditions that can lead to either of these results

**Classification of anemia:**

* Decreased Production of RBCs:
  + Iron Deficiency Anemia
  + Aplastic Anemia
* Increased Destruction of RBCs:
  + Hemolytic Anemia
  + Sickle Cell Anemia
  + Thalassemia
* Blood loss:
  + Iron Deficiency Anemia
  + Hemorrhagic Anemia
* Severety:
  + Mild: Hb 110-90 g/L
  + Moderate: Hb 89-70 g/L
  + Severe: Hb < 70 g/L
* Morphologic (describe the size of RBCs):
  + (1) normocytic,
  + (2) microcytic,
  + (3) macrocytic.
* According to the amount of hemoglobin in the cell (describe the color and Hb content of the cells):
  + (1) normochromic
  + (2) microchromic.

**Manifestations of anemia**

* Pale skin, mucous membranes, lips, nail beds, and conjunctiva
  + Waxy pallor seen in severe anemia
  + Capillary refill – no change color
* Rapid, pounding heart beat
* CNS manifestations:
  + Headache
  + Dizziness
  + Light-headedness
  + Irritability
  + Slowed thought processes, dicreases attention span
  + Apathy
  + Depression.
* Impaired healing and loss of skin elasticity
* Abdominal pain, nausea, vomiting, anorexia
* Low-grade fever

**Anemia. Assessment**

* Take health history:
  + Careful diet history to identify any deficiencies,
  + Evidence of *pica* – eating clay, ice, paste.
* Observe for manifestations of anemia:
  + Muscle weakness
  + Easy fatigability:
    - frequent resting,
    - shortness of breath,
    - poor sucking (infants)

**Nursing Intervention for Anemia :**

* Monitor vital signs, capillary refill, skin color, mucous membranes.
* Examine and document the presence of pain.
* Observation of a delay in verbal response, confusion, or restlessness
* Observe and document the presence of the cold.
* Maintain the ambient temperature to keep warm the body needs.
* Provide oxygen as needed.

**Iron Deficiency Anemia**

**Causes**

- inadequate supply of iron

- impaired absorption

- blood loss

- excessive demands for iron req’d for

growth

- inability for form Hgb

* + - * Most common anemia caused by improper iron intake

-Giving cow’s milk instead of baby formula is main culprit during the first year

**Treatment**

* + - * + Ferrous Sulfate for two months
        + Restoration takes three months

Nursing Interventions:

* Educate parents about nutrition
* Explain laboratory testing
* Teach parents proper administration of iron preparations, caution about High toxicity of iron
* Vit C increases absorption
* Milk and tea with meals reduces absorption

**Sickle Cell Anemia**

Causes: genetic transmission, 2 parents with the trait have 25% chance of having child with SCD

* + - * Autosomal recessive, sickle shaped RBCs
      * Asymptomatic in infancy prior to 5-6 months
      * Clinical course characterized by episodic crises

**Management and Nursing Care**

A- Prevent sickling by:

۞ promote tissue oxygenation and prevent tissue hypoxia through:

* + Avoid Strenuous physical activity
  + Emotional stress
  + Avoid Environments with low oxygen content

۞ promote proper hydration by:

* + Recognize signs of [dehydration](http://www.nlm.nih.gov/medlineplus/ency/article/000982.htm) and Provide access to fluids
  + Avoid excess exposure to the sun

۞ prevent infection by:

* + Keep child properly immunized as recommended by the health care provider
  + Early detection of signs and symptoms of infection
  + Frequent measured of vital signs

B- Promote supportive therapies during crises

۞ Control pain by:

* Administer analgesic and Comfortable position
  + apply warmth to painful area

۞ promote rest by keep child at bed rest

C-Replace blood and observe for signs of transfusion reactions

D- Prevent psychological problem

E-Promote long term care and genetic counseling

**Hemophilia**

Refers to group of bleeding disorders in which there deficiency of one of the factors necessary for coagulation of the blood. It get the name in 1828 by Dr. Fredrich About 80 %of all cases are demonstrated as x-linked recessive the most common forms of disorders are factors VIII classic or type A hemophilia. Other types are IX Christmas or type b, and XI type C. It is more common in males than female.

**Clinical Manifestations**

* prolonged bleeding
* Epistaxia ,bleeding in mouth ,
* GIT and spinal cord bleeding ,Petechia
* Hemoarthrosis :joint bleeding

**Medical management**   


**Nursing care:**

* Prevent bleeding
* Recognize and control bleeding (RICE)
  + Rest
  + Ice
  + Compression
  + Elevation
* Prevent crippling effects of bleeding
* Client education
* NO  rectal temperatures
* **Responsibility of nurses toward Hemarthosis**
* Observe and record the signs and symptoms of hemarthrosis such as hematoma ,local pain ,swelling limited motion and may be affixed unusable joint
* Elevate the joint and immobilized
* Active range of motion exercises
* Allow the child to control the degree of exercise according to the degree of discomfort
* Physical therapy to promote maximum function of joint an unaffected body part
* Give analgesic to control pain according Dr.
* In chronic cases my be need orthopedic interventions such as casting, application of traction or aspiration of blood
* Avoid excessive handling or weight –bearing for 48 hr.

# Thalassaemia

is a group of blood disorders. Thalassaemia is the most common inherited blood disorder. Inherited means when something is passed down from parent to child. People with thalassaemia have an increased destruction of red blood cells (hemolytic anemia). children with less red blood cannot produce enough hemoglobin. Hemoglobin is the part of the blood which carries oxygen around the body , but is more common in people of Mediterranean, Middle Eastern or Asian origin



**Types of Beta Thalassaemia**

Thalassaemia minor : children with mild thalassemia have no symptoms , sometimes called "healthy carriers of thalassaemia"

Thalassaemia Intermedia: have moderate signs and symptoms and some time need blood (occasionally)

Thalassaemia major: have signs and symptoms of thalassemia and need blood frequently

## Symptoms of Thalassemia

* symptoms of mild thalassemia.

They may feel tired or irritable; be short of breath, dizzy or lightheaded; or have pale skin, lips or nail beds compared to their normal color.

* Severe thalassemia symptoms:
* Heart palpitations
* Jaundice - yellow color in the whites of the eyes; maybe yellow tint in the skin for some skin colors
* Enlarged liver or spleen
* Enlarged bones, mainly in the cheeks and forehead
* Slowed growth - which may include later puberty - due to anemia

## 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](http://www.healthline.com/health/hemoglobin-electrophoresis).

**Management of thalassemia**

* Blood Transfusions
* Surgery splenctomy
* Bone Marrow Transplants
* Other Treatments
* Antibiotic and vaccine to help prevent infections.
* Folic acid is a B vitamin that helps build red blood cells.

**Complications of Thalassemia**

* Growth failure and pubertal delay
* Skin change and skeletal abnormalities
* Hepatosplenomgaly and Heart failer
* Endocrines abnormalities such as (insulin disorder ,hypothyroidism)
* Psychological problems to child and his family such as (daily activity ,school problem

**Nursing management responsibility toward thalassemia**

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**Leukemia**

**Causes of leukemia**

* Natural or artificial ionizing radiation
* Genetic predispositions
* Certain kinds of chemicals
* The leukemia increase with identical twins
* The leukemia associated with disorders of the immune mechanisms of the body

**signs and symptoms**

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

**Treatment and Plan of Care**

* Chemotherapy: three phases
  + Induction phase
  + Consolidation
  + Delayed intensification
  + Remission and maintenance
* Radiation
* Bone marrow transplants

**Leukemia – Diagnosis**

* History & physical
* Peripheral blood smear reveals anemia, thrombocytopenia, and *neutropenia*
* Leukemicblasts   may be seen on smear
* Bone Marrow aspiration is the definitive test
* Normal marrow contains less than 5% blasts
* Leukemic marrow has much higher percentage, often 60-100% blasts

**Leukemia – Priority Nursing Diagnoses**

* Risk for infection
* Risk for injury
* Activity intolerance
* Anxiety
* Risk for   ineffective family coping
* Pain

**Leukemia – Nursing Management**

* Prepare child/family for diagnostic tests and procedure
* Relieve pain
* Provide emotional support to child and family
* Prevent complications of myelosuppression
  + Infection
  + Private room
  + Strict hand washing
  + Restriction of visitors
  + Adequate nutrition
  + Masks
  + Hemorrhage
  + Platelet infusions
  + Avoid skin puncture
  + mouth care
  + No rectal temperatures
  + Avoid activities that could cause injury
  + General and frequent nursing assessment for detecting complications of this disease ,side effect of chemotherapy and signs and symptoms of infection
  + When blood transfusion using the nurse explain the purposes and procedure to make child feel better
  + When child has anemia the nurse keep him at rest period and balanced meals and give child adequate of protein ,CHO and fluid in order to prevent malnutrition and anemia
  + Antiemetic schedule need to be minimize or prevent the nausea and vomiting
  + Observe carefully for hemorrhage such as petechia and ecchymosis in the skin
  + Observe general hygiene such as oral hygiene and skin hygiene for any inflammation
  + Pain management
  + Recording of vital signs and body weight
  + Provide education for childُs family about diagnosis ,prognosis and treatment of leukemia
  + Provide continued emotional support for child and his family

**Blood Transfusion**

Complications:

* Hemolytic reactions
  + chills, shaking, fever
  + dyspnea
  + flank pain
  + progressive signs of shock
* Febrile reactions
* Allergic reactions
  + urticaria, flushing
  + wheezing
* Circulatory overload

**Nursing Care**

* Take VS before administering blood
* Check ID of recipient with donor’s blood type
* Check temperature of blood
* Administer with NS
* Use appropriate filter
* Observe any abnormal reaction

If reaction suspected: Stop the transfusion, maintain patent IV line with NS, take VS, notify practitioner