Blood Disorders in Children

MSc. Pediatric Nursing
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Outline

1. Introduction
2. The definition, pathophysiology , clinical Manifestation , Diagnosis, therapeutic management , nursing consideration of most common blood disorder
   A. Anemia
      - Iron deficiency anemia.
      - Sickle cell anemia
      - B-Thalasemia.
      - Aplastic anemia
   B. Hemophilia.
   C. Idiopathic thrombocytopenic purpura.
   D. Leukemia
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Introduction:

Blood is the life-maintaining fluid that circulates through the body's heart, arteries, veins, and capillaries. It carries away waste matter and carbon dioxide, and brings nourishment, electrolytes, hormones, vitamins, antibodies, heat, and oxygen to the tissues. Because the functions of blood are many and complex, there are many disorders that require clinical care by a physician ,nurse or other healthcare professional. These conditions include anemias, bleeding disorders, as well as cancers of the blood. Blood disorders are physical condition that prevents normal function of the blood in the body. A disorder may involve factors that interfere with the production of individual components found in the blood such as hemoglobin or blood proteins. The nature of blood disorder may also include situations where the blood does not coagulate properly, or the blood cells themselves are malformed or infected.

A-Anemia

Definition of Anemia

Anemia is a term used to indicate that the patient has a decrease in the number of red blood cells (RBCs) or the hemoglobin (Hb) below normal value for age (Wong's ,2013) . As a result of this decrease, the oxygen – carrying capacity of the blood is diminished, causing reduction in the oxygen available to the tissues.

Classification and Etiology of anemia's

Classification of anemia's according to Etiologic or physiology factors or Morphologic factors.

I. Etiologic factors (Wong's):

1- Impaired or decreased production of red blood cells or hemoglobin resulting from nutrition deficiency e.g .Iron deficiency lead to iron deficiency anemia e.g bone marrow failure lead to a plastic anemia.

2- Accelerated destruction of red blood cells: RBCs can be destroyed as the results of extracorpuscular defects (outside the cell) or intracorpuscular defects (inside the cell).

a- Extracorpuscular defects that cause hemolysis of normal RBC as Toxic drug, poisoning substances such as lead, thermal injury, transfusion reactions and infections as splenic enlargement.

b- Intracorpascular defect that shorten cells life span include:

- Abnormalities of Hb synthesis as sickle cell anemia and Thalasemia.
- Abnormalities of the RBC synthesis membrane and metabolism.

3- Blood loss: It may be acute or chronic e.g hemophilia

II. Morphologic classification

1- Size: microcytic [abnormally small R.B.C.s], normocytic [normal size R.B.C.s] or macrocytic [abnormally large R.B.C.s].

2- Shape: Irregularly shaped R.B.C.s, e.g. sickle cells

3- Color: Reflects Hb concentration e.g. hypochromic [abnormal decrease in Hb] or normochromic [normal red Hb color].

	Hemoglobin		**Hematocrit**		**Mean Corpuscular Volume**	
Age	**Mean**	**Low Limit**	**Mean**	**Low Limit**	**Mean**	**Low Limit**
	(g/dL)	**(g/dL)**	**(%)**	**(%)**	**(fL)**	**(fL)**
Birth	16.5	13.5	51	42	108	98
8 weeks						
Preterm	8.8	7.1	26	21		
Term	11.0	9.0	35	27		
6 months	11.0	9.5	33	28		
1 year	12.0	10.5	36	33	78	80
6 year	12.5	11.0	37	34	81	75
Adult						
Female	14.0	12.0	42	36	90	80
Male	15.5	13.5	47	41	90	80

Values derived from Nathan DG, Orkin SH, look at, Gisburg D (eds):Nathan & Infancy and childhood, 5th ed Philadelphia, WB Saunders, 1997. appendix 1,p.

Assessment criteria of Anemia:

I. General Manifestation:

- Dyspnea on exertion.
- Easy fatigability.
- Shortness of breath.
- Waxy pallor in severe anemia.
- Muscle weakness.
- Pale skin
- Poor sucking (infants).
- Tachycardia

II. Central nervous system manifestations:

- Headache.
- Dizziness.
- Night headache.
- Irritability.
- Slowed thought processes.
- Decreased attention span.
- Depression.

III. Shock (blood loss anemia):

- Poor peripheral perfusion.
- Skin is moist and cool.
- Low blood pressure.
- Increased heart rate

Therapeutic management

Treating underlining causes and makeup for any deficiency of blood ,blood component, substances to blood need for normal functioning :

- Blood cells are replaced after hemorrhage
- Nutritional anemia the specific deficiency is replaced
- In severe anemia supportive medical care may include oxygen therapy ,bed rest ,and replacement intravascular volume with intravenous (IV) fluid .

Research :

- Low Vitamin D Tied to Anemia in Healthy Children
- Dr. Meredith Atkinson of Johns Hopkins School of Medicine and her colleagues report (Journal of Pediatrics , 2013): Children with 25-hydroxyvitamin D levels below 30 mg/ml were nearly twice as likely to have anemia compared to those with normal 25(OH)D levels.

Nursing consideration(Wong's ,2013 & NANDA)

1. Parents and child should take care of the following points:
    - Nutrition(intake of iron).
    - History of chronic recurrent infection.
    - Eating habits (pica and ingestion of lead-based paint or other toxic agents.
    - Bowel habits and presence of frank blood in stools or black, tarry stools Bowel habits and presence of frank blood in stools or black, tarry stools.
    - Familial history of hereditary disease such as sickle cell disease or thalassemia.

2. Prepare child and family for laboratory test: Nurse is responsible for prepare child and family for test by :
    - Explain the significant of each test
    - Encourage parent or another supportive person to be with the child during procedure
    - Allowing the child to play with equipments on a doll or participate in the actual procedure as clean by alcohol swab
3. Decrease the child oxygen needs: So the nurse is responsible about the following:
    - To assess the child's daily living activities and play tolerance.
    - Observe vital signs behavior during periods of rests.
    - Observe vital signs and behavior during periods of activities and compare between too levels
4. An essential nursing responsibility is instruction parents in the administration of iron. Oral iron should be given in three divided doses between meals.
5. A acidify fruits or juice should be taken with medication that aids in absorption.
6. Vomiting and/or diarrhea are not uncommon complications of iron therapy. If the parents report these symptoms, the iron can be given with meals and the dosage reduced and then increased gradually.
7. Liquid preparations of iron may temporarily stain the teeth. If possible, the medication should be taken through a straw or given through a syringe or medicine dropper placed toward the back of the mouth. Brushing the teeth after administration of drug lessens the discoloration.
8. Iron preparations for I.M injection should be injected deeply into a large muscle mass using the z-tract method, and the injection site should not be massaged after injection to minimize skin staining and irritation.

9. Nurse is responsible for prepare child and family for test by :

- Explained the significant of each test
- Encourage parent or another supportive person to be with the child during procedure
- Allowing the child to play with equipment on a doll or participate in the actual procedure as clean by alcohol swab

10. Decrease tissue oxygen need by :

- Assess energy level and minimize excess demand
- Appropriate actives use is planned as music ,watching TV ,drawing , listing to stories.

11. Prevent complications :

- Hypoxia result of oxygen needed
- Prevent exposure to Infection
- Maintain adequate nutrition

A-1Iron deficiency anemia

Iron deficiency anemia is a decrease in the number of red blood cells due to a lack of iron. Iron deficiency anemia is the most common form of anemia. Iron is an essential part of hemoglobin, the oxygen-carrying protein in blood. Get iron through certain foods, and body reuses iron from old red blood cells (Glader B , 2007).

Babies are born with about 500 milligrams (mg) of iron in their bodies. By the time they reach adulthood they need to have about 5,000 mg. Children need to absorb an average of 1 mg per day of iron to keep up with the needs of their growing bodies. Since children only absorb about 10% of the iron they eat, most children need to receive 8-10 mg of iron per day. Breastfed babies need less, because iron is absorbed 3 times better when it is in breast milk(Glader B , 2007).

Causes (Wong's 2013,Glader B; 2007):

- An iron-poor diet is a common cause of iron deficiency.
- Drinking too much cow's milk is a common cause of iron deficiency in young children because cow’s milk contains little iron and can get in the way of iron absorption.
- Cow's milk also can cause problems in the intestine that lead to blood loss and increased risk of anemia.
- The adolescent growth spurt is another high-risk period.
- Iron deficiency in children can be related to lead poisoning or slow bleeding.

Symptoms (Glader B; 2007):

- Blue-tinged or very pale whites of eyes
- Blood in the stools
- Brittle nails
- Decreased appetite (especially in children)
- Fatigue
- Headache
- Irritability
- Pale skin color (pallor)
- Shortness of breath
- Sore tongue
- Unusual food cravings (called pica)
- Weakness

Note: There may be no symptoms if anemia is mild.

Exams and Tests

The health care provider will perform a physical exam. A blood sample is taken and sent to a laboratory for examination. Red blood cells appear small when looked at under a microscope.

Specific tests that may be done include:

- Hematocrit
- Serum ferritin reveals the amount of iron stored in body
- Serum iron shows how much iron is in blood
- Total iron binding capacity (TIBC) to measure the ability of a protein called transferrin to carry iron in the blood

Prognosis

With treatment, the outcome is likely to be good. In most cases, the blood counts will return to normal in 2 months. However, child should continue taking iron supplements for another 6 to 12 months or as the health care provider recommends. This will help the body rebuild its iron storage.

Possible Complications

Iron deficiency anemia can affect school performance. Low iron levels are an important cause of decreased attention span, reduced alertness, and learning difficulties, both in young children and adolescents.

Prevention

Diet is the most important way to prevent and treat iron deficiency.

Good sources of iron include:

- Apricots
- Oatmeal
- Spinach
- Tuna

Better sources of iron include:

- Chicken and other meats
- Dried beans and lentils
- Eggs
- Fish
- Peanut butter
- Soybeans

The best sources of iron include:

- Baby formula with iron
- Breast milk (the iron is very easily used by the child)
- Infant cereals and other iron-fortified cereals
- Liver

Guidelines for Iron Deficiency Management (2010) (British Columbia Medical Association ,Protocols Advisory Committee& Ministry of health services) .

I. Commonly used oral iron preparations include: ferrous gluconate, ferrous fumarate, and ferrous sulfate.

II. Iron intolerance is very common;

a. Oral iron preparations may cause nausea, vomiting, dyspepsia, constipation, diarrhea or dark stools.

b. Strategies to minimize these effects include: start at a lower dose and increase gradually over 4 to 5 days; giving divided doses or the lowest effective dose.

III. Iron absorption enhanced by taking them on an empty stomach (at least 1.5 to 2 hours after a meal), with acidic juices or vitamin C, and not with other multivitamin, calcium, or antacid tablets.

IV. Anemia will correct within 2 to 4 months if appropriate iron dosages are administered and underlying cause of iron deficiency is corrected.

V. If the patient's clinical status is compromised by moderate to severe anemia, consider admission to an acute care facility and blood transfusion. Once the patient is stable, iron replacement can be commenced.

a. Oral iron replacement is preferred to intravenous (IV) therapy when there is: inadequate iron absorption, continued blood loss, noncompliance or intolerance to oral iron therapy.

b. Complete or partial failure of monitored iron therapy trial may be due to insufficient absorption or ongoing loss (e.g. hemorrhage) or both. It should be investigated appropriately.

c. Intramuscular (IM) iron therapy is not recommended except in institutions with facility for treating anaphylactic reactions. Additional risks of IM iron therapy include unpredictable absorption and local complications (e.g. pain, staining of the skin, sarcoma formation).

VI. Recommended dose is 1 to 2 mg/kg/day of elemental iron (max 15 mg of elemental iron/day).

VII. Recommend infants and toddlers with suspected IDA begin treatment with oral ferrous sulphate.

VIII. Recommended treatment dose for infants and children is 3 to 6 mg of elemental iron/kg/day in divided doses .

Guidelines for Management of Iron Deficiency Anemia According to BritishColumbia Medical Association (2004)

Recommendation 1:

Screening is done for increased risk of iron deficiency due to physiological factors, clinical suspicion and multiple risk factors.

Recommendation 2:

Encourage individuals to consume diet with sufficient iron. Foods contain iron in two forms:

- Heme "iron" is present in red meat, fish and poultry and its absorption is not dependent on factors present in food.
- Non-Heme iron is present in fruits, vegetables, cereals and dairy products and its absorption is affected by other factors.

Recommendation 3:

- Tea, coffee, herbal teas and cocoa taken within one hour of meal inhibit iron absorption.
- Vitamin C (strawberries, tomato, citrus fruit) enhance iron absorption.

Researches:

- **Iron Supplements Improve Cognition and Health in Anemic Kids**

  Daily iron supplements improve cognitive and physical health in primary school children with anemia have better cognitive and physical health when given daily iron supplements, according to a systematic review and meta-analysis published online October 15 in the *Canadian Medical Association Journal*.

- **Iron Deficiency Linked to Psychiatric Disorders in Kids**

  Children and adolescents with iron deficiency anemia (IDA) are at increased risk for psychiatric disorders, including depressive disorder, bipolar disorder, anxiety disorder, and autism, new research shows by *BMC Psychiatry,2013*

Nursing consideration (Wong's ,2013 & NANDA)

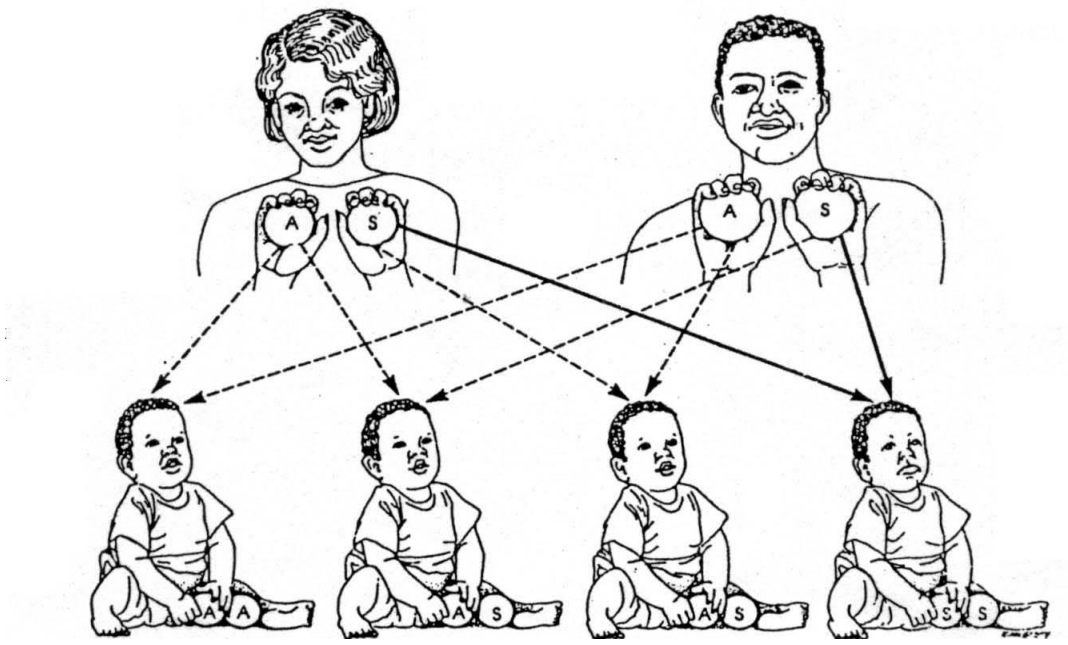
Essential responsibility of nurse is instruction parent about iron as prescribed

- Oral iron divided doses in between meals because to decrease hydrochloride acid for absorption .
- A citrus fruit or juice aid in absorption oral iron .

- Cow's milk or milk products contain substance bind and interfere with oral iron absorption (Carley ,2003) .
- Liquid iron may stain the teeth ,so may given in syringe and placed toward the back of the mouth. Also ,brushing the teeth after administration of the drug .
- If parental iron is prescribed ,iron dextran must be injected deeply in large muscle in Z- track method .the site not massage to minimize skin straining and irritation .
- They should IV route should be considered adverse reaction ,as anaphylaxis and test done is recommended before routine use.
- Parent should educated about breast milk is poor in iron sources after 5 months of lactation so supplementation in exclusively breast fed infant to 6 month of age.
- The nurse discusses with parent importance of using iron fortified formula .
- Diet education of teenagers is difficult ,especially because teenage girls are particularly prone to following weight –reduction diets
- Explain the potential adverse effects of iron which includes nausea and vomiting, diarrhea or constipation or black stools and tooth discoloration.
- Instruct care givers to keep iron supplements out of reach of children since it is toxic when overdosed.

A-2 Sickle Cell Anemia

It is one of a group of disease collectively termed hemoglobinopathies, in which normal adult hemoglobin A (Hb A) is partly or completely replaced by abnormal sickle hemoglobin (HbS) Wong's , 2013).



Pathophysiology (Wong's):

The clinical features of SCA are primary the result of:

1- Abnormal obstruction caused by the sickled RBCs.

2- Increased RBCs destruction.

The entanglement of rigid sickle shaped cells with one another intermittently block the microcirculatarculation, causing vaso-occlusion .the resultant absences of blood flow to adjacent tissue causes local hypoxia ,leading to tissue ischemia and infarction (cellular death).Most of the complications seen in SCA can be traced to this process and its impact on various organs of the body.

Clinical manifestations (Wong's ,2013) :

Manifestations do not appear in the first 3-4 months of life as Hb F protects against sickling.

Two groups of manifestation:

1- Manifestations of chronic moderately severe hemolytic anemia that is not routinely transfusion –lead to in general Possible growth retardation ,chronic

anemia level 6-9g/dl, possible delayed sexual maturation and marked susceptility to sepsis .

2- Episode crises of 4 types :

- **Vaso-occlussive crises** or painful episode characterized by distal ischemia and pain . Clinical manifestations are caused by ischemia resulting from severe hemolytic anemia and chronic vaso-occlusive episodes :
    - ✶ Heart: murmur, cardiac decompansation, heart failure.
    - ✶ Lung: pneumonia & emboli in pulmonary vessels by occluded mass of sickled R.B.C.s.
    - ✶ Spleen: splenomegaly, decreased phagocytic function which leads to infection and rupture of enlarged spleen ⇢ internal hemorrhage.
    - ✶ Liver: hepatomegaly, chronic production of bilirubin ⇢ jaundice and gallstone.
    - ✶ Kidney: inability to concentrate urine, hematuria, polyuria & enuresis.
    - ✶ Central nervous system (CNS): headache, vision disturbance, aphesia, convulsion, stroke hemiplegia & death.
    - ✶ Skeletal muscles: symmetrical painful swelling of hands & feet (hand – foot syndrome) during infancy. Kyphosis may occur and chronic hypoxia ⇢osteomyelitis.
- **Sequestration crisis** ,a pooling of blood in the liver and spleen with decreased blood volume and shock.
- **Aplastic crisis** ,diminished RBC production resulting in profound anemia.
- **Hyperhemolytic crisis** , an accelerated rate of RBC destruction characterized by anemia ,jaundice and reticulocytosis .

Diagnostic evaluation:

- Hemoglobin electrophoresis to detect abnormal Hb.

- Sickle-turbidity test (sickledex) anticoagulated blood is mixed with special solution, Since Hgbs is normally much less soluble than adult Hb (HgbA) or fetal Hb (HgbF), when it mixed with this solution, and it forms cloudy or turbid mixture.
- Serum bilirubin.
- Complete blood picture.
- Liver function test.
- Sedimentation rate.
- X-ray for skull and long bone.

Therapeutic management: (Wong's,2013)

1-Rest to minimize energy expenditure and oxygen use.

2-Hydration through oral and intravenous therapy.

3-Electrolyte replacement because hypoxia results in metabolic acidosis.

4-Analgesics for severe abdominal and joint pain from vaso- occlusion.

5-Blood replacement to treat anemia.

6-Antibiotics to treat any existing infection.

7-Administration of pneumococcal and meningococcal vaccines is recommended for these children because of their susceptibility to infection as a result of a functional asplenia.

Guidelines for Management of Sickle Cell Disease in Children according to CLINICAL PRACTICE (2004)

A. Acute Painful Episode (Vaso Occlusive Disease)

For mild to moderate pain give:

1) Acetaminophen (15 mg/kg/dose, maximum dose 65 mg/kg/day,) with codeine (1 mg/kg/dose, maximum dose 60 mg/dose and 6 mg/kg/day). These can be given separately or together.

2) For moderate to severe pain (or if pain relief as given above for mild to moderate pain is inadequate after 30-60 minutes), give an IV bolus of: Morphine 0.1-0.15 mg/kg/dose (maximum 7.5 mg/dose). Repeat morphine once after 60 minutes if pain relief inadequate. Administer a 10 mL/kg bolus of normal saline IV, followed by 1.5 times the maintenance fluid requirement of D5W with 0.45 NaCl IV

For severe pain:

1) Continue Morphine Infusion

2) Additional IV boluses of morphine 0.05 mg/kg can be given q 1-2 h prn.

3) If adequate pain relief is established for 2 hours with 1 or 2 doses of intermittent IV morphine, consider administering acetaminophen with codeine .

4) B. Fever (Rule Out Infection)

1) If the culture is positive and the organism is penicillin-susceptible, change to IV penicillin (250,000 units/kg/day, divided q4-6h, maximum 20 x 106 units/day)

2) If the culture is positive for penicillin-non-susceptible pneumococcus, ensure that the patient is on vancomycin, in addition to ceftriaxone/cefotaxime .

3) If the patient has penicillin/cephalosporin-resistant pneumococcal meningitis, or is not improving after 36-48 h of therapy, do a repeat lumbar puncture .

Nursing consideration (Wong's ,2013& NANDA):

1. Monitor vital signs. Assess for pain.
2. Obtain blood and urine culture, chest x-ray and CBC results if infection is the cause of sickling.

3. Monitor child's nutritional intake with hydroxyurea. If taken orally, this drug can cause anorexia.
4. Assess for kidney function by noting if the child has urinated or not. (Kidney infarction may occur)
5. Do not administer potassium if the kidney function is not verified. Potassium if not excreted by the kidney may cause arrhythmia.
6. Educate family and child
   - Seek early intervention for problem ,as fever
   - Give penicillin as ordered
   - Recognize signs and symptoms of splenic sequestration as well as respiratory problems that can lead to hypoxia
   - Treat child normally, but can get sick in way that other children cannot.
7. Promote supportive therapies during crisis
   - Any pain program should combine with psychological support to help child deal with depression , anxiety & fear.
   - Heat to affected area is soothing .Cold compress are not applied to the area because enhance sickling and vasoconstriction .
8. Support the family (discuss their family)
9. If family member have the SCD trait or SCA genetic counseling is important because 25% chance with each pregnancy of having child with the disease

Research :

- **Influenza Vaccine Safe for Kids with Sickle Cell Anemia**

- Children with sickle cell anemia who receive the trivalent inactivated influenza vaccine are no more likely to be hospitalized for sickle cell crisis than children with sickle cell anemia who do not receive the vaccination, according to a study published online of *Pediatrics* ,2012. The study is important because children with sickle cell anemia have a 56-fold greater risk for hospitalization for influenza complications compared with children without sickle cell anemia.

A-3 β-Thalassemia [Cooley's anemia]

Thalassemia is an inherited disorder characterized by deficient in rate of production of specific globing chain in Hgb (Wong's ,2013) .

Thalassemia is a blood disorder in which the body makes an abnormal form of hemoglobin. This can happen when there is an imbalance in the different protein components that make up a hemoglobin molecule. Thalassemia is an inherited disorder and more common in people of Mediterranean, Middle Eastern, Chinese and Southeast Asian ancestry.

Types of thalassemia

Four protein chains make up hemoglobin—two alpha globin and two beta globin chains. There are two major types of thalassemia -- **alpha thalassemia** and **beta thalassemia** -- named after defects that can occur in these protein chains.

Alpha Thalassemia

Four genes, two from each parent, are required to make alpha globin protein chains. When one or more genes are missing, it produces alpha thalassemia. Alplha thalassemia occurs frequently among Southeast Asians, Middle East Asians, Chinese and Africans.

Beta Thalassemia

Beta thalassemia major is also called Cooley's anemia. There are normally two beta globin genes, one from each parent. Beta thalassemia is a change in one or both of the beta globin genes. This chart describes the different types of beta thalassemia. Beta Thalassemia occur frequently to those of

Mediterranean origin and lesser to Chinese, other Asians and African Americans.

Affected beta genes	Disorder	Anemia symptoms	Other names
1	silent carrier	mild	Beta thalassemia minor
1	trait	mild	
2	intermedia	moderate	
2	major	severe	Cooley's anemia

Pathophysiology

Beta thalassemias are due to mutations in the HBB gene on chromosome 11, also inherited in an autosomal-recessive fashion. The severity of the disease depends on the nature of the mutation. Mutations are characterized as either β^o or β thalassemia major if they prevent any formation of β chains, the most severe form of β thalassemia. Also, they are characterized as β^+ or β thalassemia intermedia if they allow some β chain formation to occur. In either case, there is a relative excess of α chains, but these do not form tetramers: Rather, they bind to the red blood cell membranes, producing membrane damage, and at high concentrations they form toxic aggregates.

At birth the baby with thalassemia major seems entirely normal. This is because the predominant hemoglobin at birth is still fetal hemoglobin (Hb F). Hb F has two alpha chains (like Hb A) and two gamma chains (unlike Hb A). It has no beta chains so the baby is protected at birth from the effects of thalassemia major.

Anemia begins to develop within the first months after birth. It becomes progressively more and more severe. The infant fails to thrive (to grow normally) and often has problems feeding (due to easy fatigue from lack of oxygen, with the profound anemia), bouts of fever (due to infections to which the severe anemia predisposes the child) and diarrhea and other intestinal problems.

Clinical manifestations

- Clinical manifestations of Thalassemia major are due to presence of **hemosiderine**, an insoluble form of iron stored in tissues.

- The excessive **hemosiderine** leads to **hemosiderosis** [excessive iron storage in tissues without tissue damage} and **hemochromatosis** {excessive iron storage in tissue with tissue damage].

- **Clinical manifestations include :**

1- Anemia (before diagnosis):

- pallor
- Unexplained fever.
- Poor feeding.
- Markedly enlarged spleen.

2-With progressive anemia:

Signs of hypoxia:

- Headache.
- Bone pain.
- Decreased exercise tolerance.
- Restlessness.
- Anorexia.

3-Other features:

- Small stature
- Delayed sexual maturation.

- Bronzed, freckled complexion

4-Bone changes (older children if untreated):

- Enlarged head.
- Flat or depressed bridge of the nose.
- Enlarged maxilla.
- Generalized skeletal osteoporosis

Diagnostic evaluation:

1- Hb electrophoresis HgbA and HgbF levels confirm diagnosis.

2- Reticulocyte count, serum iron level, total iron-binding capacity.

3- Structurally impaired R.B.C.s and has short life span.

4- Low Hb and hematocrit level.

5-Radiographs of involved bones reveal characteristic findings.

Therapeutic management (Wong's ,2013):

The objective of supportive therapy is to maintain sufficient Hgb levels to prevent bone marrow expansion and the resulting bony deformities, and to provide sufficient RBCs to support normal growth and development so:

- **Blood transfusion** is the foundation of the medical management with the goals to maintain the Hgb level above 9.5g/dl .**The advantage of the therapy include :**

1-Improved physical and physiological well-being because it increases the ability to participate in normal activities.

2- It decreased cardiomegaly and hepatomegaly.

3-Fewer bone changes.

4-Near normal growth & development until puberty.

5-Fewer infections.

- **Chelation therapy.**

One of the potential complications of frequent blood transfusions is iron overload. Because the body has no effective means of eliminating the excess iron, the mineral is deposited in the body tissues. To minimize development of hemosiderosis, Desferal iron chelating therapy which causes exertion of more iron in urine & coloring it orange red is given with oral vitamin C.

- **Desferal** given in a dose 20 to 40 mg /kg by slow subcutaneous infusion via infusion pump over 8-12 hours / days / week.
- **Folic acid supplementation**
- **Splenectomy**

Italian Society of Hematology Guidelines for the Management of Iron Overload in Thalassemia Major (2008)

Recommendation 1:

- Patients with known transfusion history need to have serum ferritin level determined 1-2months to determine value of iron load to be used for initiating iron chelating therapy.

- Patients over 5 years of age with unknown transfusion history should have both serum ferritin and liver iron concentration determined in order to plan iron chelating therapy.

Recommendation 2:

- Iron chelating should be started after children have received more than 10 units of blood or with serum ferritin level over 1000 mg/mL.

- In patients with unknown transfusion history iron chelating should be started when liver iron content is over normal range.

Recommendation 3:

- Children, who start iron chelation therapy before 6 years of age, when body iron burden is modest, and in whom goal of chelation therapy is prophylactic maintenance of iron balance, should receive desferal.

Recommendation 4:

- Patients undergoing iron chelation should receive periodic monitoring of serum ferritin.

- Liver iron content should be assessed in order to avoid under- or over-treatment.

- Repeat liver iron content every year during chelation therapy.

Recommendation 5:

- Patients with evidence of non-compliance to deferoxamine, or with severe adverse effects from deferoxamine without existing severe iron overload, oral iron chelator should be used

- Patients who develop severe iron overload (serum ferritin higher than 3,000 ng/mL maintained for three months, liver iron content higher than 15 mg/g) or iron-related cardiomyopathy should receive "intensive" or "combined" iron chelation therapy. The first choice for combined therapy is desferal associated with deferiprone.

Standards of Care Guidelines for Thalassemia (**2012**) **Published by**Children's Hospital & Research Center Oakland supported by Centers for Disease Control (CDC) & Thalassemia Support Foundation (TSF)

Recommendations for care

- Folic acid supplements and avoidance of oxidative compounds and medications are recommended.

- At routine visits, growth, development, facial bone deformity, dental status, and hepatosplenomegaly should be monitored.

- Routine monitoring of hemoglobin levels is required.

- Patients with hemoglobin H disorders develop neonatal anemia. Splenomegaly and hypersplenism are relatively common. Splenectomy usually ameliorates the severe anemia noted in nondeletional hemoglobin H cases. Splenectomy may be required at a very young age in transfusion-dependent cases. Prophylactic antibiotics and infection precautions are similar to other splenectomy

patients. Thrombosis prevention is indicated in cases requiring splenectomy. Low-dose aspirin or other anticoagulants may be used.

- Ongoing monitoring of iron stores with quantitative imaging of the liver is indicated because of the unreliability of serum ferritin tests.
- In nontransfused patients, imaging should be initiated in early adolescence.
- Cardiac function monitoring is indicated.
- Bone-density measurement should be initiated in early adolescence.

Nursing consideration

- Promote compliance with transfusion and chelating therapy
- Assist child in coping with anxiety provoking treatments and the effect of illness
- Foster the family and child adjustment to a chronic illness
- Observe for complications of multiple blood transfusion.

A-4 Aplastic Anemia "AA":

Refere to bone marrow failure condition in which formed elements of the blood are simultaneously depressed .The peripheral blood smear demonstrates pancytopenia.

Etiology:

This condition can be congenital or acquired, and it is more common between ages 15-25 years old.

Common causes of Acquired A plastic Anemia

- Infection with human parvovirus (HPV) or over whelming infection
- Irradiation
- Drug such as the chemotherapeutic agents and several antibiotics

- Industrial and household chemicals including benzene and its derivatives which are found in petroleum products ,dyes ,paint remover

- Infiltration and replacement of myeloid elements such as in leukemia or the lymphomas

- Idiopathic in which no identifiable cause can be found

Clinical Manifestation:

- In congenital AA, clinical symptoms can include: purpura, petechiae (pinpoint lesions), bleeding, fatigue, and pallor. Laboratory findings include: neutropenia (decreased number of neutrophils), thrombocytopenia (low platelets count) that progress to pancytopenia (decreased number of blood cell components). Children here are at risk of developing malignancies such as acute non-lymphocytic leukemia.

- In acquired AA, clinical manifestations are related to the degree of bone marrow failure can include: purpura, petechiae, bleeding, pallor, weakness, tachycardia, and fatigue.

- Bone marrow aspiration and biopsy must be performed. In normal bone marrow, 40% to 60% of the marrow space is typically occupied with hematopoietic cells (depending on the age of the person) by contrast, the bone marrow in patients with aplastic anemia typically contains very few hematopoietic cells and consists primarily of fatty space and stromal cells

Guidelines for the diagnosis and management of aplastic anemia (2009)Blackwell Publishing Ltd, British Journal of Haematology, 147, 43–70

This guideline on the diagnosis and management of a plastic anemia was published in 2003 in the British Journal of Hematology. This guideline is an update of the 2003 guideline and is to replace the 2003 guideline.

A. Investigations required for diagnosis a plastic anemia

1. Full blood count, reticulocyte count, blood film and % HbFIn most cases the haemoglobin level, neutrophil and platelet counts are all uniformly depressed, but in the early stages isolated cytopenia, particularly thrombocytopenia, may occur.
2. Bone marrow examination
3. Definition of disease severity based on the FBC and bone marrow findings
4. Liver function tests and viral studies Liver function tests should be performed to detect antecedent hepatitis, but in post-hepatitic aplastic anemia the serology is most often negative for all the known hepatitis viruses.
5. **Vitamin B12 and folate levels** Vitamin B12 and folate levels should be measured to exclude megaloblastic anemia which, when severe, can present with pancytopenia.
6. **Autoantibody screen**
   The occurrence of pancytopenia in systemic lupus erythematosus may (i) be autoimmune in nature occurring with a cellular bone marrow or (ii) be associated with myelofibrosis or rarely (iii) occur with a hypocellular bone marrow. Blood should be tested for anti-nuclear antibody and anti-DNA antibody in all patients presenting with aplastic anemia.
7. **Tests to detect a PNH clone**
   Paroxysmal nocturnal haemoglobinuria should be excluded by performing flow cytometry (Dacie & Lewis, 2001; Parker et al, 2005). The Ham test and sucrose lysis test have been abandoned by most centers as diagnostic tests for PNH. Analysis of The occurrence of pancytopenia in systemic lupus erythematosus may (i) be autoimmune in nature occurring with a cellular bone marrow or (ii) be associated with myelofibrosis or rarely (iii) occur with a hypocellular bone marrow.
8. Trial therapy or clinical research protocols
9. Management of aplastic anemia in the presence of an abnormal cytogenetic clone
10. Management of patients with aplastic anemia who have a significant PNH clone, resulting in clinical and/or laboratory evidence of haemolysis
11. Management of aplastic anemia in pregnancy

B. Management of aplastic anemia

Recommendations

i. Prophylactic platelet transfusions should be given when the platelet count is <10 · 109/l (or <20 · 109/l in the presence of fever).

ii. Irradiated blood products should be given routinely to all patients having ATG treatment.

iii. Transfusion of irradiated granulocyte transfusions may be considered in patients with life-threatening neutropenic sepsis.

iv. The routine use of **RECOMBINANT HUMAN ERYTHROPOIETIN** rHuEpo in aplastic anemia is not recommended.

v. A short course of **Granulocyte colony-stimulating factor** (**G-CSF** or **GCSF**) may be considered for severe systemic infection that is not responding to intravenous antibiotics and anti-fungal drugs, but should be discontinued after 1 week if there is no increase in the neutrophil count.

vi. Prophylactic antibiotic and antifungal drugs should be given to patients with neutrophil count <0.2 $X10^9$/l.

vii. Infection or uncontrolled bleeding should be treated first before giving immunosuppressive therapy. This also applies to patients scheduled for BMT, although it may sometimes be necessary to proceed straight to BMT in the presence of severe infection as a BMT may offer the best chance of early neutrophil recovery.

viii. Haemopoietic growth factors, such as rHuEpo or GCSF, should not be used on their own in newly diagnosed patients in an attempt to 'treat' the aplastic anemia.

Nursing management

- Monitor nutritional intake and output

- Monitor weight gain corporate clients Explain to klie about the importance of nutrition for the body and the diet prescribed and ask again what has been described
- Help clients and families identify and select foods that contain calories and protein in accordance with a programmed diet.
- Serve food in warm and attractive
- Collaboration with a dietitian to determine the proper diet as well as physicians in the provision of vitamins. Monitor vital signs
- Review the causes of intolerance client activity
- Train ROM when circumstances allow clients
- Teach client techniques energy savings for activity
- Increase client activity according to ability Monitor vital sign and signs of infection
- Monitor laboratory results (number of leucocytes and differential)
- Perform aseptic techniques and Septic each perform action on the client.
- Observations on the region where the stabbing infusion, catheter
- Teach the client and families about how infection prevention and signs of infection

2- Hemophilia

Hemophilia refers to a group of bleeding disorders in which there is deficiency of one of the factors necessary in which there is a deficiency of one of the factors necessary for coagulation of the blood. (Wings, 2008) Hemophilia A is most common 90% than hemophilia B . Hemophilia A occurs in about 1 in every 5,000-10,000 male births .18,000 people in America have Hemophilia . Four hundred babies are born with hemophilia each year.

Hemophilia is caused by a deficiency of coagulation factor VIII (FVIII) (hemophilia A) or factor IX (FIX) (hemophilia B) related to

mutations of the clotting factor gene. The life expectancy of persons born with hemophilia, who have access to adequate treatment, is approach normal with currently available treatment.

Pathophysiology

The basic defect of hemophilia A is deficiency of factor VIII (ant hemophilic factor (AHF). AHF is produced by the liver and is necessary for the formation of thromboplastin in phase I of blood coagulation .The less AHF found in the blood , the more severe the disease . Individual with hemophilia have two of the three factors required for coagulation: vascular influence and platelets.
Therefore they may bleed for longer period, but not at a faster rate.

Diagnosis of Hemophilia

Accurate diagnosis is important and essential for effective management.

- History as Easy bruising in early childhood, Spontaneous bleeding (particularly into the joints and soft tissue).
- Complete blood cell count
- Coagulation studies
- FVIII assay
- Hemoglobin/hematocrit
- Platelet count
- Bleeding time and prothrombin time.
- Activated partial thromboplastin time (aPTT)
- Imaging studies
- CT & MRI for head and spinal column are used for further assessment of spontaneous or traumatic hemorrhage
- Ultrasonography is useful in the evaluation of joints affected by acute or chronic effusions.
- Testing for inhibitors are indicated when bleeding is not controlled after adequate amounts of factor concentrate are infused during a bleeding episode. Inhibitor concentration is titrated using the Bethesda method, as follows:

- Positive result: Over 0.6 Bethesda units (BU)
- Low-titer inhibitor: Up to 5 BU
- High-titer inhibitor: Over 5 BU

Clinical Manifestation (Wong's, 2013)

- Prolonged bleeding anywhere from or in the body
- Subcutaneous and intramuscular hemorrhage
- Hemarthrosis (bleeding in the joint space)
- Intracranial hemorrhage
- Retroperitoneal hemorrhage
- Postsurgical bleeding,
- Excessive bruising ,easy bruising (eccymosis)

Guidelines for the management of hemophilia ,The National Guideline Clearinghouse (2013)

Management/Treatment

1. General care and management:
    - Principles of care (treating acute bleeds as quickly as possible, easily accessible patient identification with information about the bleeding disorder and treatment used, use of desmopressin [DDAVP] for mild-moderate hemophilia)
    - Coordinated comprehensive care team
    - Encouragement of physical fitness and activity while protecting target joints
    - Prophylactic factor replacement therapy
    - Regular monitoring of health status and outcome
    - Management of pain due to chronic hemophilic arthropathy (avoiding aspirin and non-steroidal anti-inflammatory drugs [NSAIDs])
    - Performing surgery at or in consultation with a comprehensive hemophilia treatment centre

2. Special management issues:
    - Testing clotting factor levels of potential carriers (e.g., female relatives of hemophilia patient)
    - Genetic testing for carrier status of at-risk female family members of people with hemophilia
    - Chorionic villus sampling (CVS) or biopsy for prenatal diagnosis of hemophilia
    - Delivery of infants with known or suspected hemophilia
    - Vaccinations, including immunization to hepatitis A and B
    - Considerations for care of hemophilia patients with comorbid conditions
3. Hemostatic agents: viral-inactivated plasma-derived or recombinant clotting factor concentrates (factor VIII, factor IX), cryoprecipitate, fresh frozen plasma, DDAVP, tranexamic acid, epsilon aminocaproic acid
4. Treatment of specific hemorrhages: joint, muscle, throat and neck, gastrointestinal, acute abdominal, ophthalmic, renal, oral, soft tissue, and central nervous system hemorrhages; head trauma; epistaxis; and lacerations and abrasions
5. Management of complications of hemophilia:
    - Management of musculoskeletal complications
    - Assessment and quantification of inhibitor levels and management of complications related to inhibitors
    - Management of transfusion-transmitted and other infection-related complications including human immunodeficiency virus (HIV), hepatitis B, and hepatitis C

Nursing consideration

Prevent bleeding

- By appropriate exercise to strengthen muscles and joint and to allow age appropriate activity.
- During infancy and toddler hood the normal acquisition of motor skills creates innumerable opportunities for falls ,bruises ,minor wound
- Environment safe

- School nurse should plan appropriate schedule of activity with safe equipment.
- To prevent oral bleeding, softening tooth brush in warm water before use to minimize trauma to the gum.
- Avoid IM injection as possible.
- No aspirin should be used

Recognize and control bleeding

- Apply **RICE** (Rest - Ice - Compression - Elevation) for immediate bleeding.

Support family and prepare for home care

- Genetic counseling after diagnosis
- Children who have become infected with HIV through transfusion and factor replacement products are faced with the consequences of this dreaded disease.

3- Idiopathic Thrombocytopenic Purport (ITP):

Idiopathic thrombocytopenic purport (ITP) Or Immune thrombocytopenic purport is acquired hemorrhage disorder characterized by:

1) Thrombocytopenia, excessive destruction of platelet
2) Purport ,a discoloration caused by petechiae the skin
3) A normal bone marrow with normal increased number of immature platelet (Megakaryocytic) and eosinophils .

Etiology:

It has been suggested that, in some cases, ITP may result from

(i) T cell-mediated cytotoxicity or

(ii) antibody-mediated complement activation causing platelet lysis or

(iii) antibodymediated suppression of megakaryocyte production (Nakhoul

et al, 2006).

Most often this is a result of antibody production against platelets, though in a small number of cases a type of white blood cell called T-cells will directly attack platelets. This immune system error may be a result of the following:

- Infections, typically viral infections, including the viruses that cause chicken pox, hepatitis C, and AIDS, can prompt antibodies that cross-react with platelets.
- Immune disorders, such as rheumatoid arthritis and lupus
- Low-grade lymphomas and leukemias may produce abnormal antibodies against platelet proteins

Clinical Manifestation:

- Skin: petechia , purpura or ecchymosis (more common in the anterior surface of the leg and over bony prominence as ribs ,scapulae, shoulders and pubic area)
- Mucous membranes :conjunctiva ,buccal mucosa ,gums ,soft palate ,nose (epistaxis)
- Hematuria and GIT bleeding may occur
- CNS bleeding (rare &serious) lead to headache ,convulsion ,coma
- Spleen not usually palpable (its tip is felt in ‹ 10 % of cases)

Treatment:

According to royal children hospital: Clinical Practice Guidelines Management,2012

Children fall broadly into two categories:

1. Acute (~ 90%): self limiting disease (sometimes preceded by a viral syndrome) with spontaneous resolution within 6 months (usually within 2 months).
2. Chronic (~ 10%): does not remit within 6 months.

Treatment as an Outpatient

Most patients with a platelet count > 20 x 10^9/l and some of those with a platelet count < 20 x 10^9/l can be managed as outpatients with no specific treatment.

Treatment as an Inpatient

1. Any patient with ITP who has active bleeding (oral, aural, nasal, rectal, etc) even if resolved should be admitted and considered for oral prednisolone (2-4 mg/kg/day for 2 weeks then tapered). Normal human immunoglobulin is also effective but is not usually used for initial treatment
2. Avoid aspirin and non-steroidal anti-inflammatory drugs

British Journal of Hematology 2003

Recommendations:

1. Children with chronic ITP usually do not need active therapy but should be followed up regularly and reminded to report to hospital after injuries. They should have a designated contact person and number.
2. Children with chronic severe ITP should be referred to a pediatric hematologist for management and long term follow-up.

Guideline on the Evaluation and Management of Immune Thrombocytopenia (ITP) , 2011 Clinical Practice

Recommendation

• A single dose of IVIg (0.8-1.0 g/kg) or a short course of corticosteroids should be used as first-line treatment.

• IVIg should be used instead of corticosteroids if a more rapid increase in platelet count is required.

• There is no evidence to support using corticosteroids for longer courses compared to very brief courses.

• Anti-D may be considered for first-line therapy in Rh+ non-splenectomized children

• In children, splenectomy or other interventions with potentially serious complications should be delayed for at least 12 months, unless warranted by severe disease unresponsive to other measures or due to quality of life considerations.

• If previous treatment with corticosteroids, IVIg, or anti-D has been successful, these options may be used as needed to prevent bleeding.

• If previous treatment with corticosteroids, IVIg, or anti-D has been unsuccessful, subsequent treatment may include splenectomy, rituximab, thrombopoietin receptor

MMR-related ITP:

• Children with a history of ITP who are not immunized should receive their scheduled first MMR vaccine.

• In children with either non-vaccine or vaccine-related ITP who have already received their first dose of MMR vaccine, vaccine titers can be checked. If the child displays full immunity, no further MMR vaccine should be given. If the child does not have adequate immunity, then the child should be re-immunized at the recommended age.

Nursing consideration :

Nursing care focuses on controlling and reducing the number of bleeding episodes. Assess vital signs and level of consciousness. Abdomen is assessed for hepatosplenomegaly. Preventing measure are similar to those of hemophilia. The skills needed to monitor IV infusion and blood transfusion and to administer heparin are the same as for any child receiving theses therapies.

4-Leukemia

Leukemia is a malignant disease of blood forming organs of the body that results in uncontrolled growth of immature white blood cells (WBCs). It can be defined as a group of malignant disorders characterized by arrest of

maturation and uncontrolled proliferation of precursors of white blood cells in the bone marrow.

Leukemia is the most common form of cancer in children followed by brain tumors and lymphomas. It accounts for about one third of pediatric malignancies. The annual incidence is 3 to 4 cases per 100,000 white children younger than 16 years of age (Wong's ,2008) .it occurs more frequently in males than in females after age one year ,and the peak onset is between 2 and 6 years of age. Untreated leukemia results in death from infections or hemorrhage within 6 months of affection.

Classification of leukaemia

1) Acute lymphatic leukaemia (ALL)

Usually occurs before 14 years of age peak incidence is between 2-9 years of age. It arising from a single lymphoid stem cell, with impaired maturation and accumulation of the malignant cells in the bone marrow.

2) Acute Myelogenous Leukaemia (AML)

It occurs at any age but occurs most often at adolescence. Characterized by the development of immature myeloblasts in the bone marrow.

Etiology

The exact etiology of acute leukemia is unknown, but it results from interaction of a number of risk factors e.g.: Combination of predisposing factors including genetic and environmental influences, exposure to ionizing radiation, Chemicals such as benzene, insecticides and pesticides, electro magnetic fields (EMF), drug intake especially alkylating agents and, accidentally X-ray and drug exposure especially during first trimester of pregnancy ..

Pathophysiology

In leukemia, the bone marrow creates an overabundance of abnormal young white cells (blasts). “Blast” is a short name for an immature white blood cell such as lymphoblast, myeloblast, or monoblast. Normally, less than 5 percent of the blast cells are present in healthy bone marrow at any time. It develops into mature functioning white blood cells and are not usually found in the bloodstream. Leukemic blasts remain immature, multiply continuously, provide no defense against infection and may be present in large numbers in the bloodstream and bone marrow. After accumulating in the bone marrow, it deprives the normal cells of the essential nutrients for metabolism lead to decrease RBCs, PLT, WBCs production. leukemic cells can spill over into the blood and cross the blood-brain barrier and invade the central nervous system (brain and spinal cord).

Clinical manifestations of leukemia

The 3 main consequences are: **(1**) anemia from decreased erythrocytes**, (2)** infections from neutropenia, **(3)** bleeding from decrease platelet production.

The first symptoms of ALL in children usually are pallor, low-grade fever, and lethargy as a result from anemia (caused by decreased RBCs production). The child may easily have petechiae and bleeding from oral mucous membranes bruise because of a low platelets count (thrombocytopenia). These symptoms may develop gradually or may be sudden in onset. As the diseases progress, the spleen and liver begin to enlarge due to infiltration of abnormal cells leading to abdominal pain, vomiting and anorexia. Bone and joint pain occurs when abnormal lymphocytes invade the bone periosteum and center nervous system (CNS) manifestation. Signs or symptoms of CNS involvement are rarely observed at the time of the initial diagnosis. The signs and symptoms include headache, nausea, vomiting, lethargy, irritability, nuchal rigidity and papilledema. Cranial nerve involvement may occur which most frequently involves the third, fourth, sixth, and seventh cranial nerves. Leukemia can present as intracranial or spinal mass, which causes numerous neurologic symptoms, most of which are due to nerve compression. Physical assessment reveals painless generalized lymphadenopathy especially of the submandibular or cervical nodes .Acute Myelogenous Leukaemia (AML) Similar to ALL plus sternal tenderness.

Diagnosis of leukemia

Leukemia is usually suspected by history, physical manifestations and a peripheral blood smear that contains immature forms of leukocytes, frequently combined with low blood counts. Definitive diagnosis is based on flow cytometry of the cells obtained by biopsy or bone marrow aspiration of cerebrospinal fluid (CSF) examination which identifies the specific type of blast cell. After the diagnosis is confirmed, a lumber puncture is performed to determine if there is any CNS involvement. CNS involvement occurs when there is more than five blasts / cmm .

An articles zinc deficiency in lymphoma and leukemia Mentioned that With regard to antioxidant role of zinc in the body, zinc deficiency may be considered as one of the factors for development and progression of cancer.

Complications of leukemia

Complication of leukemia includes metastasis to the blood, bone, CNS and liver or other organs and alteration in growth. A late complication includes problems with neuron-cognitive function, ocular, cardiovascular or thyroid dysfunction.

Guideline on management of leukemia (American Society of Clinical Oncology (ASCO),2013)

Three types of treatments are used to treat childhood ALL: chemotherapy, radiation treatment, and stem cell transplantation/bone marrow transplantation. Sometimes, these treatments are used in combination. Treatment options and recommendations depend on several factors, including possible side effects, and the patient's preferences and overall health.

1) **Chemotherapy** :Chemotherapy is the use of drugs to kill cancer cells, usually by stopping the cancer cells' ability to grow and divide. Systemic chemotherapy is delivered through the bloodstream to reach cancer cells throughout the body.

Chemotherapy is the primary treatment for ALL. It may be given by mouth (orally), injected into a vein or muscle, or injected into the cerebral spinal fluid (CSF). It is generally done in four phases:

A. **Remission induction therapy** uses chemotherapy to kill as many of the leukemia cells as possible to cause the cancer to go into remission.

B. **Central nervous system directed therapy** kills any leukemia cells in the central nervous system and prevents the spread of the disease to the spinal fluid.

C. **Consolidation therapy** begins when the child's leukemia has gone into remission. Higher doses of chemotherapy are used to kill the majority of the remaining leukemia cells.

D. **Continuation or maintenance therapy** lasts for two to three years to kill any remaining (residual) leukemia cells.

The side effects of chemotherapy depend on the individual and the dose used, but they can include short-term side effects like hair loss, fatigue, loss of appetite, nausea and vomiting, diarrhea, and kidney and liver dysfunction. These side effects usually go away once treatment is finished. Other side effects related to chemotherapy that may last longer or develop after treatment is finished include bone and joint problems and learning problems. The severity of the side effects depends on:

- The type and amount of the drug being given and the length of time the child receives the drug.
- Child experiences and age

2) **Radiation therapy**

Radiation therapy is the use of high-energy x-rays or other particles to kill cancer cells. The most common type of radiation treatment is called external-beam radiation therapy, which is radiation given from a machine outside the body. A radiation therapy regimen (schedule) usually consists of a specific number of treatments given over a set period of time.

Radiation therapy for ALL is generally used only when the leukemia has spread to the brain, spinal fluid, or a boy's testicles, or in high-risk disease

to help prevent the spread of leukemia to the spinal fluid. Radiation therapy is more often used for patients with T-cell leukemia.

Side effects from radiation therapy may include hair loss, fatigue, mild skin reactions, upset stomach, and loose bowel movements. Most side effects go away soon after treatment is finished. However, long-term side effects of radiation treatment to the brain and body can occur and may possibly include hormone problems affecting growth and metabolism, learning problems, and an increased risk of developing a second cancer including a brain tumor. Skin, salivary gland, and thyroid cancers can also occur after treatment for ALL .

3) **Stem cell transplantation/bone marrow transplantation**

Stem cell transplantation is most often used as a treatment for recurrent or refractory ALL. Rarely, transplantation may be recommended as part of the initial therapy when leukemia is associated with very high risk features. A stem cell transplant is a medical procedure in which diseased bone marrow is replaced by highly specialized cells, called hematopoietic stem cells. Hematopoietic stem cells are found both in the bloodstream and in the bone marrow. Today, this procedure is more commonly called a stem cell transplant, rather than bone marrow transplant, because it is the blood stem cells that are typically being transplanted, not the actual bone marrow tissue.

Before recommending transplantation, doctors will talk with the patient and family members about the risks of this treatment and consider several other factors, such as the type of cancer, results of any previous treatment, and patient's age and general health.

There are two types of stem cell transplantation depending on the source of the replacement blood stem cells: allogeneic (ALLO) and autologous (AUTO). In both types, the goal of transplantation is to prevent the cancer cells in the marrow, blood, and other parts of the body from returning. The transplant allows the patient's marrow to be replaced with healthy blood stem cells from another source. In most stem cell transplants, the patient is treated with high doses of chemotherapy and/or radiation therapy to destroy as many cancer cells as possible. **Palliative/supportive care**

Leukemia and its treatment often cause side effects. In addition to treatment to slow, stop, or eliminate the disease, an important part of care is relieving a child's symptoms and side effects. This approach is called palliative or supportive care, and it includes supporting the patient with his or her physical, emotional, and social needs.

Palliative care can help a child at any stage of illness. Children often receive treatment for the leukemia and treatment to ease side effects at the same time. In fact, patients who receive both often have less severe symptoms, better quality of life, and report they are more satisfied with treatment.

Recurrent ALL

If the ALL recurs, treatment depends on many factors, including the type of treatment the child received originally, the length of time between the initial diagnosis and the recurrence, and whether leukemia cells are found in the bone marrow, CSF, testicles, or in more than one of these sites when it recurs.

When ALL recurs, patients and their families often experience emotions such as disbelief or fear. Families are encouraged to talk with their health care team about these feelings and ask about support services to help them cope.

If treatment fails

Although treatment is successful for the majority of children with cancer, sometimes it is not. If a child's leukemia cannot be cured or controlled, this is called advanced or terminal leukemia. This diagnosis is stressful, and it may be difficult to discuss. However, it is important to have open and honest conversations with the child's doctor and health care team to express the family's feelings, preferences, and concerns. The health care team is there to help, and many team members have special skills, experience, and knowledge to support patients and their families.

Parents or guardians are encouraged to think about where the child would be most comfortable: at home, in a home-like setting elsewhere, in the hospital, or in a hospice environment. Nursing care and special equipment can make staying at home a workable alternative for many families. Some children may be happier if they can arrange to attend school

part-time or keep up other activities and social connections. The child's health care team can help parents or guardians decide on an appropriate level of activity. Making sure a child is physically comfortable and free from pain is extremely important as part of end-of-life care.

The death of a child is an enormous tragedy, and families may need support to help them cope with the loss. Pediatric cancer centers must have professional staff and support groups to help with the process of grieving.

Nursing consideration :

- Prepare child and family for diagnosis and therapeutic procedure

  Child must undergo several tests as bone marrow aspiration or biopsy and lumberpuncture .Multible finger stick and veinpuncture for blood analysis and drug infusion are common occurrence. Therefore the child needs an explanation of each procedure and what can be expected.also, unconscious sedation and nonpharmacologies strategies are used to reduce discomfort associated with these painful proceures.

- Relive pain: The effectiveness use of analgesia is especially important when the malignant process uncontrolled and causes acute pain .

- Prevent complications of myelosuppression: The reduce numbers of blood cells results in secondary problems of infection ,bleeding tendencies ,and anemia .

- Use precautions in administering and handling chemotherapeutic agent for example educating the family about drugs.

- Manage problems of drug toxicity :Nausea vomiting ,anorexia ,mucosal ulceration Hemorrhagic cystitis

References:

1. Hockenberry M, Wilson D, Winkelstein M. Wong's Essentials of Pediatric Nursing. 9th ed. St. Louis: Mosby Inc., 2013
2. Wu AC, Lesperance L, Bernstein H. Screening for iron deficiency. *Pediatr-Rev.* May 2002;23:171-178.
3. Glader B. Iron-deficiency anemia. In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF, eds. *Nelson Textbook of Pediatrics*. 18th ed. Philadelphia, Pa: Saunders Elsevier; 2007:chap 455.
4. **Standards of Care Guidelines for Thalassemia** (**2012**) **Published by**Children's Hospital & Research Center Oakland supported by Centers for Disease Control (CDC) & Thalassemia Support Foundation (TSF) available at web site : http://hemonc.cho. org/thalassemia/documents /SOC Guidelines 2012. pdf
5. British Journal of HaematologyVolume 147, Issue 1, Article first published online: 10 AUG 2009Guidelines for the diagnosis and management of aplastic Anaemia ,2009 Blackwell Publishing Ltd, British Journal of Haematology, available in website : http://onlinelibrary.wiley.com/doi/10.1111/j.1365 -2141.200 9.07842.x/pdf
6. Evidence for Childhood Cancers (Leukemia) 2012 , available in website : www.bioinitiative.org/.../sec 12_2012_Evidenc e_%20Ch...
7. https://client.myoptumhealth.com/myoptumhealth/guest/page.esync?view=prelogin.learn.learnLanding&command=DiseasesAndConditions&article=8cab991ac6259110VgnVCM1000005220720a____
8. http://www.chw.org/display/PPF/DocID/21844/router.asp
9. Helmy E, Omar M, Assem H, Adel M, Tawfik M, Dawood M. Madkour A's Essentials of Pediatrics. 10th ed. Alexandria: Ayad Press., 2009; 129-53
10. Sickle Cell Disease (SCD) Available at: http://www.cdc.gov/NCBDDD/sicklecell/recommendations.html.
11. BetaThalassemia Available at : http://www. medicinenet. com/beta_ thalassemia /article.htm .
12. Beta Thalassemia (Cooley's Anemia) in Children Available at : http://www.urmc.rochester.edu/encyclopedia/content.aspx?ContentTypeID=90&Con tentID=P023302012
13. Richards M, Williams M, ChalmersA E .United Kingdom Haemophilia Centre Doctors' Organization guideline approved by the British Committee for Standards in Haematology: guideline on the use of prophylactic factor VIII concentrate in children and adults with severe haemophilia . Available at http://www.slideshare.net/AnaestHSNZ/guidelines-mng-hemophilia

14. JOSEPH J. IRWIN, M.D, JEFFREY T. KIRCHNER, D.O Anemia in Children .AMERICAN FAMILY PHYSICIAN. 2001 64(8):1379- 86 AMERICAN ACADEMY OF PEDIATRICS. Health Supervision for Children with Sickle Cell Disease .2002;109;526-535 available at : http// www.pediatrics.org

15. Advisory committee. Investigation and Management of Iron Deficiency.2004availableathttp://www.nal.usda.gov/fnic/foodcomp/Data/SR14/wtrank/sr14a303.pdf

16. Angelucci E, Barosi G, Camaschella C et al. Italian Society of Hematology practice guidelines for the management of iron overload in thalassemia major and related disorders. haematologica .2008; 93(5): 741-52
17. Price V et al. A practical approach to hemophilia care in children. Paediatr Child Health 2007; 12(5): 381-3.
18. WORLD FEDERATION OF HEMOPHILIA. GUIDELINES FOR THE MANAGEMENT OF HEMOPHILIA. 2005
19. GUIDELINES FOR THE INVESTIGATION AND MANAGEMENT OF IDIOPATHIC THROMBOCYTOPENIC PURPURA IN ADULTS, CHILDREN AND IN PREGNANCY. British Journal of Hematology 2003; 120: 574–96
20. Ball J, Bindler R. Pediatric Nursing: Caring for Children. 4th Edition. Pearson 2008, Washington.
21. Hemophilia web site available on : http://emedicine.medscape.com/article/779322-overview
22. British Journal of HaematologyVolume 141, Issue 2, Article first published online: 3 MAR 2008
23. ITP web site available on : http://www.rch.org.au/clinicalguide/guideline_index/Immune_Thrombocytopenic_Purpura/
24. An articles zinc deficiency in lymphoma and leukemia (puplished in Jundishapur Sci Med J 2012;11(4)),available on website : http://journals.ajums.ac.ir/components4.php?rQV=8BkMApDZJV 2 Z V 3Zu FGb fZGfAB DQ6QnchR3cfZGf ABDNApDZJRnb lJXYw9lZ 8B0N4MDQ6QWStVGdp9lZ8BEOApDZJxWY0J3bQxWYuJXdvp2X QyATNApDZJ52b pR3Yh9lZ