The child with health problems related to the blood and its nursing care

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ANEMIA

- The term **anemia** describes a condition in which the number of red blood cells (RBCs) or the hemoglobin (Hgb or Hb) concentration is reduced below normal values for age. This diminishes the oxygen- carrying capacity of the blood, causing a reduction in the oxygen available to the tissues.

- Anemia is the most common hematologic disorder of infancy and childhood and is not a disease itself but an indication or manifestation of an underlying pathologic process.

NURSING TIP The following are suggested explanations for teaching children about blood components:

Red blood cells—Carry the oxygen you breathe from your lungs to all parts of your body.

White blood cells—Help keep germs from causing infection
Platelets—Small parts of cells that help make bleeding stop by forming a clot (scab) over the hurt area

Plasma—The liquid portion of blood, which has clotting factors that help make bleeding stop

- **Anemias are classified in relation to :**

- (1) **etiology** or **physiology**, manifested by erythrocyte or Hgb depletion, and
- (2) **morphology**, the characteristic changes in RBC size, shape, or color

- **RED BLOOD CELL MORPHOLOGY**
- **Size**
- **Variation in RBC sizes (anisocytosis)**
  - Normocytes (normal cell size)
  - Microcytes (smaller than normal cell size)
- - Macrocytes (larger than normal cell size)

- **Shape**
- **Variation in RBC shapes (poikilocytosis)**
  - Spherocytes (globular cells)
  - Drepanocytes (sickle-shaped cells)
  - Numerous other irregularly shaped cells

- **Color**
- **Variation in hemoglobin concentration in the RBC**
  - Normochromic (sufficient or normal amount of hemoglobin per RBC)
  - Hypochromic (reduced amount of hemoglobin per RBC)
  - Hyperchromic (increased amount of hemoglobin per RBC)

Consequences of Anemia

- The basic physiologic defect caused by anemia is:
- a decrease in the oxygen-carrying capacity of blood and consequently a reduction in the amount of oxygen available to the cells.
- Growth retardation, resulting from decreased cellular metabolism and coexisting anorexia, is a common finding in chronic severe anemia and is frequently accompanied by delayed sexual maturation in the older child.

Diagnostic Evaluation of Anemia

Depending one **history and physical examination**,

- lack of energy
- easy fatigability
- Pallor
- the first clue to the disorder may be alterations in the CBC,
- Although anemia is sometimes defined as an Hgb level below 10 or 11 g/dl, this arbitrary cutoff is inappropriate for all children because Hgb levels normally vary with age.

- **Therapeutic Management**

- Treat the underlying cause and to make up for any deficiency of **blood, blood component, or substance the blood needs for normal functioning.**

- In patients with severe anemia, **supportive medical care** may include:
- oxygen therapy,
- bed rest,
- replacement of intravascular volume with intravenous (IV) fluids.

- **The prognosis** for anemia depends on the correction of the cause.

- **Nursing Care Management**
- The assessment of anemia includes the basic techniques that are applicable to any condition.
- **The age of the infant or child provides some clues** regarding the **possible etiology of the anemia.**
- For example, **iron-deficiency anemia occurs** more frequently in toddlers between **12 and 36 months of age** ، and during the growth spurt of **adolescence.**
- **Racial or ethnic background is significant.**
- For example, the anemias related to abnormal Hgb levels are found in Southeast Asians and persons of African or Mediterranean ancestry. اسلاف
- These same groups may be genetically **deficient in the enzyme lactase** after the period of infancy. Affected individuals are unable to tolerate lactose in the diet, with consequent intestinal irritation and chronic blood loss.

- **Nursing Care Management**
- **Prepare the Child and Family for Laboratory Tests**
- **Decrease Tissue Oxygen Needs**
- **Prevent Complications**

Iron-deficiency Anemia

Iron-deficiency Anemia caused by an inadequate supply of dietary iron .

The prevalence of iron-deficiency anemia has decreased during infancy in the related to provides of iron-fortified formula for the first year of life and routine screening of Hgb levels during early childhood .

Preterm infants are especially at risk because of their reduced fetal iron supply.

Children 12 to 36 months of age are at risk for anemia as a result of primarily cow milk intake and not eating an adequate amount of iron-containing food .

Adolescents are also at risk because of their rapid growth rate combined with poor eating habits, menses, obesity, or strenuous activities.

- **Pathophysiology Of Iron-deficiency Anemia**
- Iron-deficiency anemia can be caused by any number of factors that decrease the supply of iron, impair its absorption, increase the body's need for iron, or affect the synthesis of Hgb.
- During the last trimester of pregnancy, iron is transferred from the mother to the fetus. Most of the iron is stored in the circulating erythrocytes of the fetus, with the remainder stored in the fetal liver, spleen, and bone marrow.
- These iron stores are usually adequate for the first 5 to 6 months in a full-term infant but for only 2 to 3 months in preterm infants and multiple births.

- **Pathophysiology of Iron-deficiency Anemia**
- not supplied to meet the infant's growth demands after the fetal iron stores are depleted, iron-deficiency anemia results. Physiologic anemia should not be confused with iron-deficiency anemia resulting from nutri- tional causes.
- Although most toddlers with iron-deficiency anemia are under-weight, many infants are overweight because of excessive milk inges- tion (known as **milk babies**).
- These children become anemic for two reasons: milk, a poor source of iron, is given almost to the exclusion of solid foods, and 50% of iron-deficient infants fed cow's milk have an increased fecal loss of blood.

- **Therapeutic Management**
- **increasing the amount of supplemental iron** the child receives.
- In **formula-fed infants**, the most convenient and best sources of supplemental iron are iron-fortified commercial formula and iron-fortified infant cereal. Iron-fortified formula provides a relatively constant and predictable amount of iron and is not associated with an increased incidence of gastrointestinal (GI) symptoms, such as colic, diarrhea, or constipation.
- If dietary sources of iron cannot replace body stores, oral iron supplements are prescribed for approximately 3 months.

• Cont: Therapeutic Management

- **Ferrous iron, more readily absorbed than ferric iron**, results in higher Hgb levels. Ascorbic acid (vitamin C) appears to facilitate absorption of iron and may be given as vitamin C–enriched foods and juices with the iron preparation.
- **If the Hgb level fails to rise after 1 month of oral therapy,** it is important to assess for persistent bleeding, iron malabsorption, non- compliance, improper iron administration, or other causes of the anemia.
- **Parenteral (IV or intramuscular [IM]) iron** administration is safe and effective but painful, expensive, and occasionally associated with regional lymphadenopathy, transient arthralgias or serious allergic reaction .
- Therefore, parenteral iron is reserved for children who have iron malabsorption or chronic hemoglobinuria.
- **Transfusions** are indicated for the most severe anemia and in cases of serious infection, cardiac dysfunction, or surgical emergency when anesthesia is required. Packed RBCs (2–3 ml/kg), not whole blood, are used to minimize the chance of circulatory overload.
- Supplemental oxygen is administered when tissue hypoxia is severe.

- **Prognosis**
- The prognosis for a child with this condition is very good. However, some evidence indicates that if the iron-deficiency anemia is severe and longstanding, cognitive, behavioral, and motor impairment may result

- **Nursing Care Management**
- An essential nursing responsibility is **instructing parents in the administration of iron.**
- Oral iron should be given as prescribed in two divided doses between meals, when the presence of free hydrochloric acid is greatest, because more iron is absorbed in the acidic environment of the upper GI tract.
- A citrus fruit or juice taken with the medication aids in absorption.
- An adequate dosage of oral iron turns the **stools a tarry green** color. The nurse advises parents of this normally expected change and inquires about its occurrence on follow-up visits.
- Absence of the greenish black stool may be a clue to poor administration of iron, either in schedule or in dosage. Vomiting or diarrhea can occur with iron therapy. If the parents report these symptoms, the iron can be given with meals and the dosage reduced and then gradually increased until tolerated.

Cont: Nursing Care Management

- **If parenteral iron preparations are prescribed**, iron dextran must be injected **deeply into a large muscle mass using the Z-track method**. The injection site **is *not* massaged** after injection to minimize skin staining and irritation. Because no more than 1 ml should be given in one site, the IV route should be considered to avoid multiple injections.
- Careful observation is required because of the risk of adverse reactions, such as anaphylaxis, with IV administration.

Cont: Nursing Care Management

- **Diet**
- A primary nursing objective is to prevent nutritional anemia through family education. Because breast milk is a low iron source, the nurse must reinforce the importance of administering iron supplementation to exclusively breastfed infants by 4 months of age
- The AAP recommends that preterm, marginally low and low–birth-weight infants, or infants with inadequate iron stores at birth receive iron supplements at approximately 2 months of age .
- In formula-fed infants, the nurse discusses with parents the importance of using iron-fortified formula and of introducing solid foods at the appropriate age during the first year of life.

QUALITY PATIENT OUTCOMES:
Iron Deficiency Anemia

- Early recognition of signs and symptoms of iron deficiency anemia
- Appropriate quantity of milk, use of iron-fortified infant formula, and introduction of solid foods
- Adherence to oral iron supplement and appropriate administration
- Hemoglobin increase within 1 month and anemia resolved within 6 months

Sickle Cell Anemia

- **What's the difference between sickle cell anemia and sickle cell disease?**

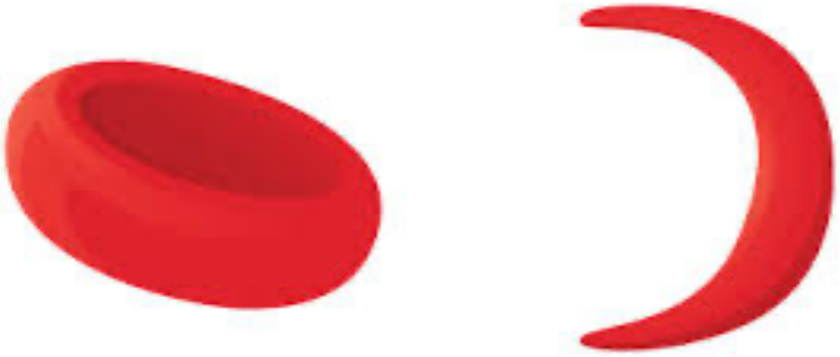
- **Sickle cell disease** is an umbrella term for the many different **types of sickle cell disorders.**
- Healthcare providers reserve the term **"sickle cell anemia**" for **the types of SCD that cause the most severe anemia. These types are hemoglobin SS and hemoglobin beta zero thalassemia.**

- The total number of SCD patients in Iraq in 2015, **was 5,124.**

- Epidemiological studies have revealed that sickle cell disease patients are clustered in two geographical areas in Iraq, one among the Arabs in the extreme south, another among the Kurdish population in the extreme north, where they constitute major health problems.

SICKLE CELL ANEMIA

- Sickle cell anemia (SCA)is one of a group of diseases collectively termed hemoglobinopathies in which normal adult Hgb (Hgb A [HbA]) is partly or completely replaced by abnormal sickle Hgb (HbS).



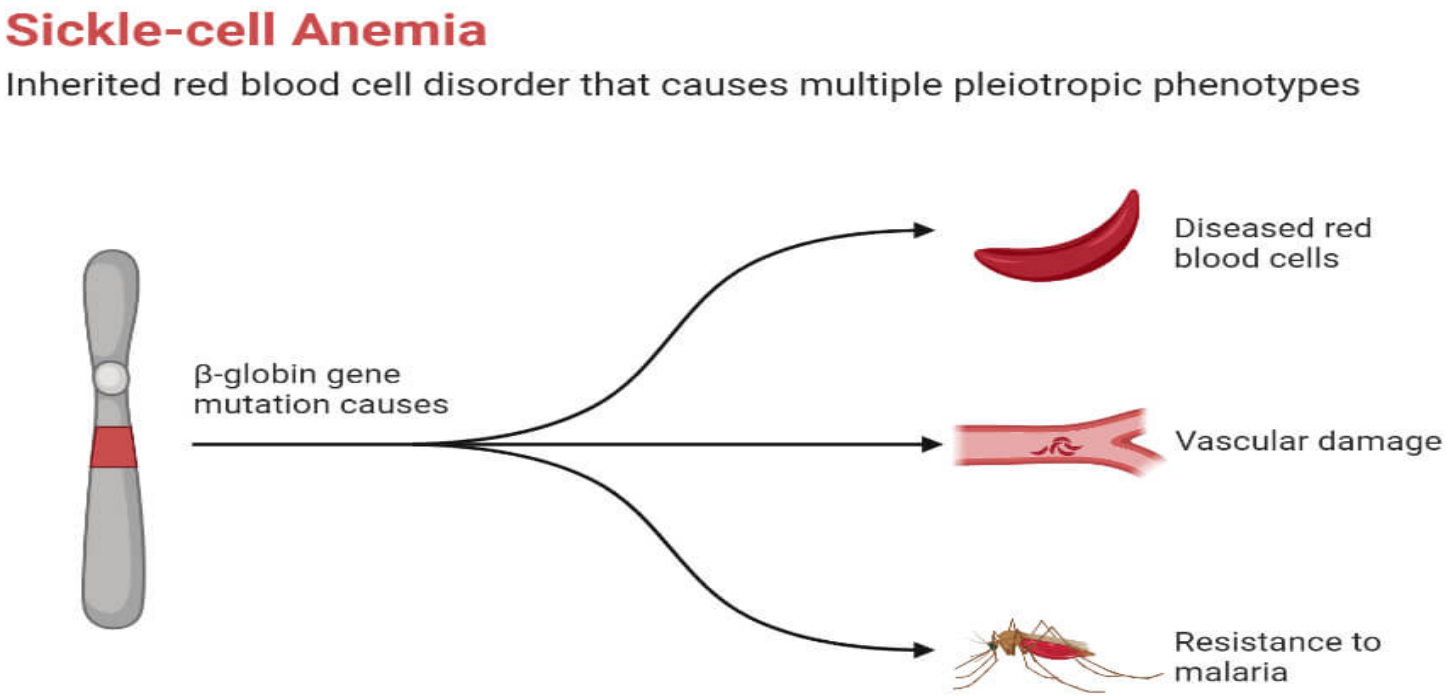
Normal Red Blood Cell

Sickle Cell

Hemoglobin

Protein iron
compound in the blood that carries oxygen to the cells and car ries carbon dioxide away from the cells.

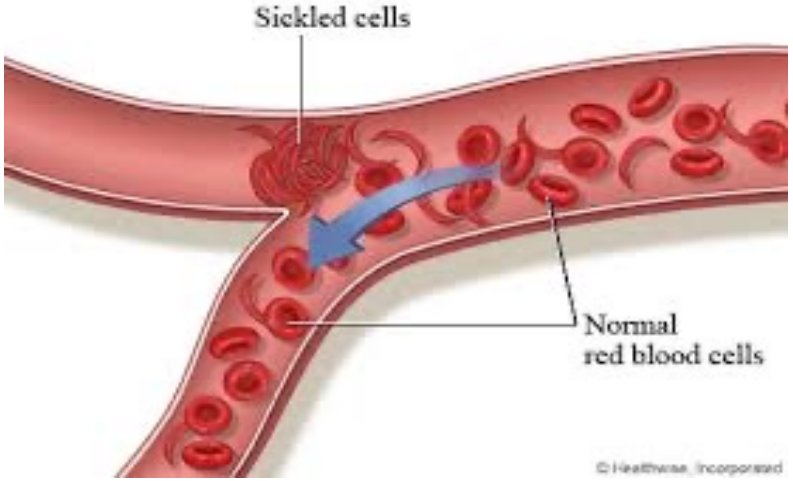
- Sickle cell disease (SCD) includes all those hereditary disorders whose clinical, hematologic, and pathologic features are related to the presence of HbS.
- Other correct terms for SCA are SS and homozygous SCD.
- It is the condition when the body produces abnormal hemoglobin, hemoglobin S, due to a mutation in the beta-globin gene ***HBB***



Sickle-cell-anemia

Pathophysiology

- The clinical features of SCA are primarily the result of
- (1) obstruction caused by the sickled RBCs,
- (2) vascular inflammation, and
- (3) increased RBC destruction.



Functions of Hemoglobin

1. Oxygen Transport

When O_2 is bound with Hgb, it is called ***oxyhemoglobin*** and Hgb without bound O_2 is called ***deoxyhemoglobin***.

2. Carbon dioxide Transport

When CO_2 is bound with Hgb, it is called ***carbaminohemoglobin***.

Other Gases/Ion Transport

Besides O_2 and CO_2, Hgb can also bind with other ligands like carbon monoxide (CO), nitric oxide (NO), sulfur monoxide (SO), nitrite ion (NO^{-2}), sulfides (S^{-2}), etc.

4. Regulation of Blood pH and Buffering Function

Hgb molecules can also bind to the hydrogen ions and maintain the pH of the blood.

- **Types of Hemoglobin**
- Based on the non-alpha subunits, normal hemoglobin is mainly of three types:

1. **Haemoglobin A**

- It is the predominant type of hemoglobin accounting for about 95 to 98% of total adult hemoglobin. It contains two alpha subunits and two beta subunits.

2. **Hemoglobin A2**

- It accounts for about 2 to 3% of total adult hemoglobin. It contains two alpha subunits and two gamma subunits.

3. **Hemoglobin F**

- It is the hemoglobin of fetuses and newborns and it is present in scanty amounts, below 1%, in adults. It contains two alpha subunits and two delta subunits.
- Besides these major hemoglobin, there are other mutated forms also like **hemoglobin E**, **hemoglobin S**, and **hemoglobin C**.

Normal Hemoglobin Level

The amount of hemoglobin in blood is expressed in grams per deciliter (g/dl).

The amount of hemoglobin depends on the age, sex, and health status of an individual.

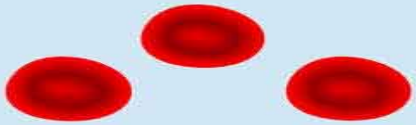
In general, the Hgb level in a human range from 12 to 20+ g/dl.

Age of Person	Normal Hgb Level (g/dl)
Newborn	14 to 24
2 weeks	13 to 20
3 months	9.5 to 14.5
6 months to 6 years	10.5 to 14.0
6 years to 12 years	11 to 16
Adult male	14 to 18
Adult female	12 to 16

Diagnostic Evaluation

- Newborn screening for SCA infants can be identified before symptoms occur.
- At birth, infants have up to 80% of HbF, which does not carry the defect.

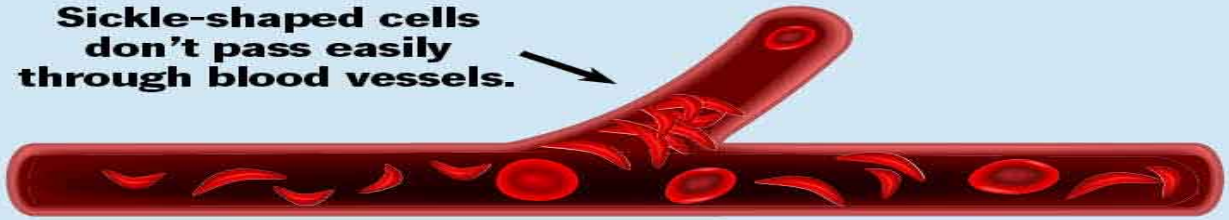
Sickle cell disease



Normal red blood cells are round and flexible.

In sickle cell disease, red blood cells stiffen, changing shape into sickles (crescent-shaped).

Sickle-shaped cells don't pass easily through blood vessels.



Sickle cell disease symptoms include:

Frequent pain episodes.

Pain affects your child's chest, back, legs and arms most often.

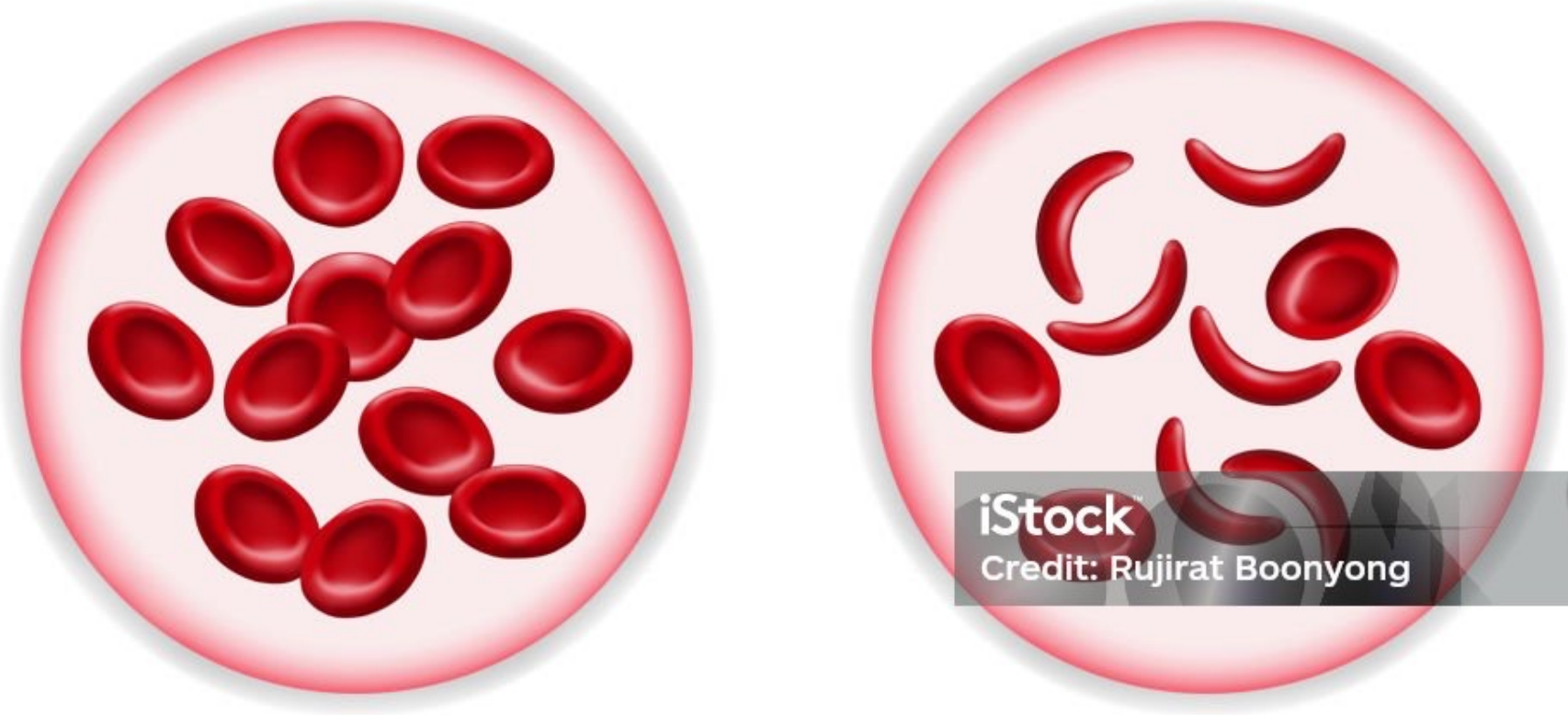
Swelling and inflammation of their joints.

Painful swelling of their hands and feet.

Anemia, causing fatigue, paleness and weakness.

Jaundice (yellowing of skin and whites of eyes).

Sickle cell anemia



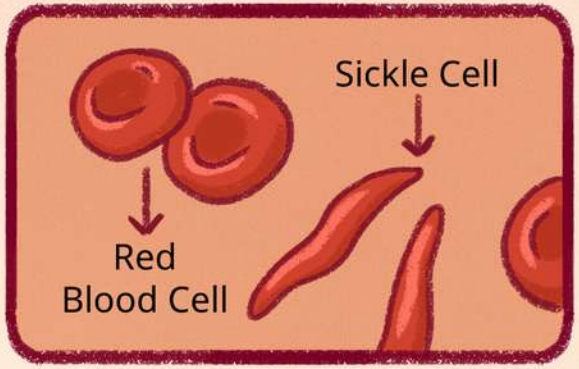
Normal red blood cells

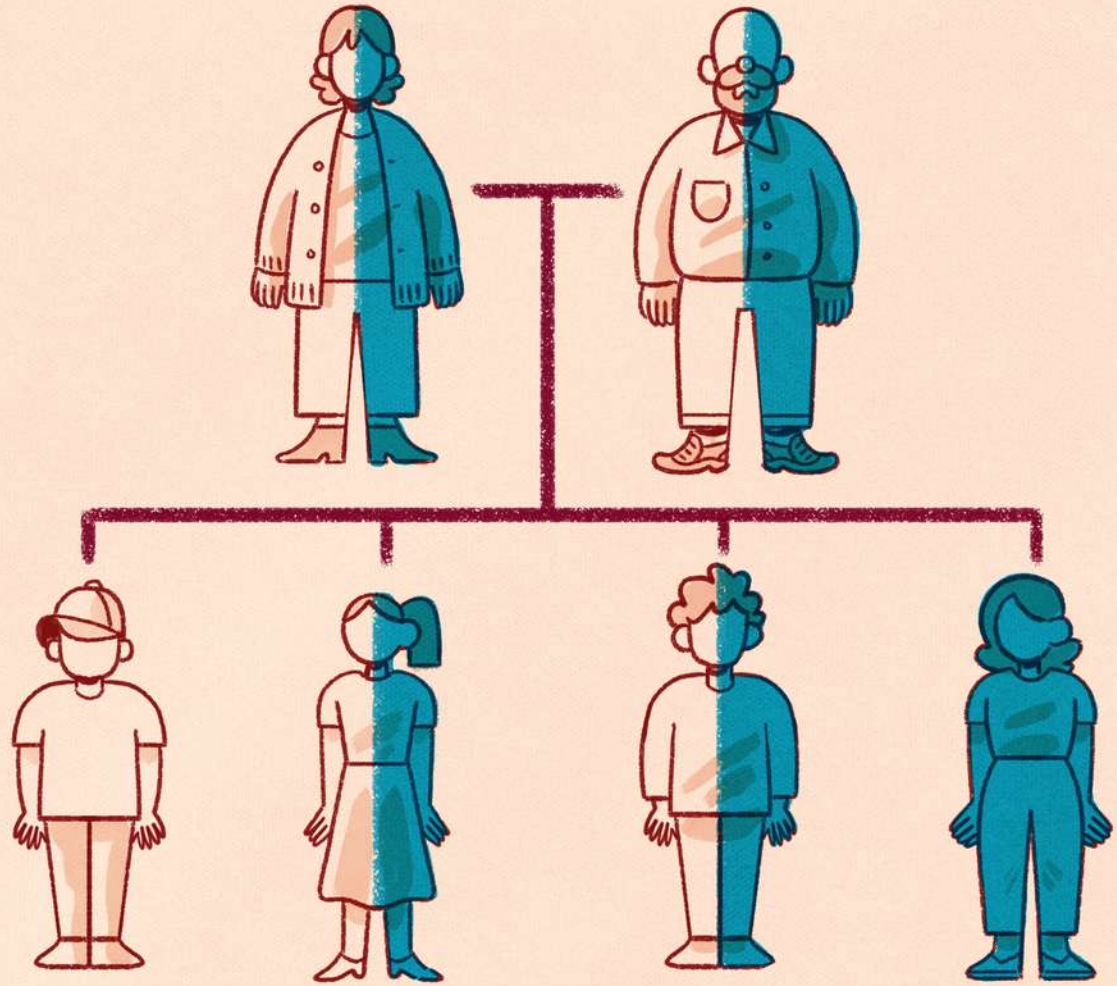
Sickle cell anemia

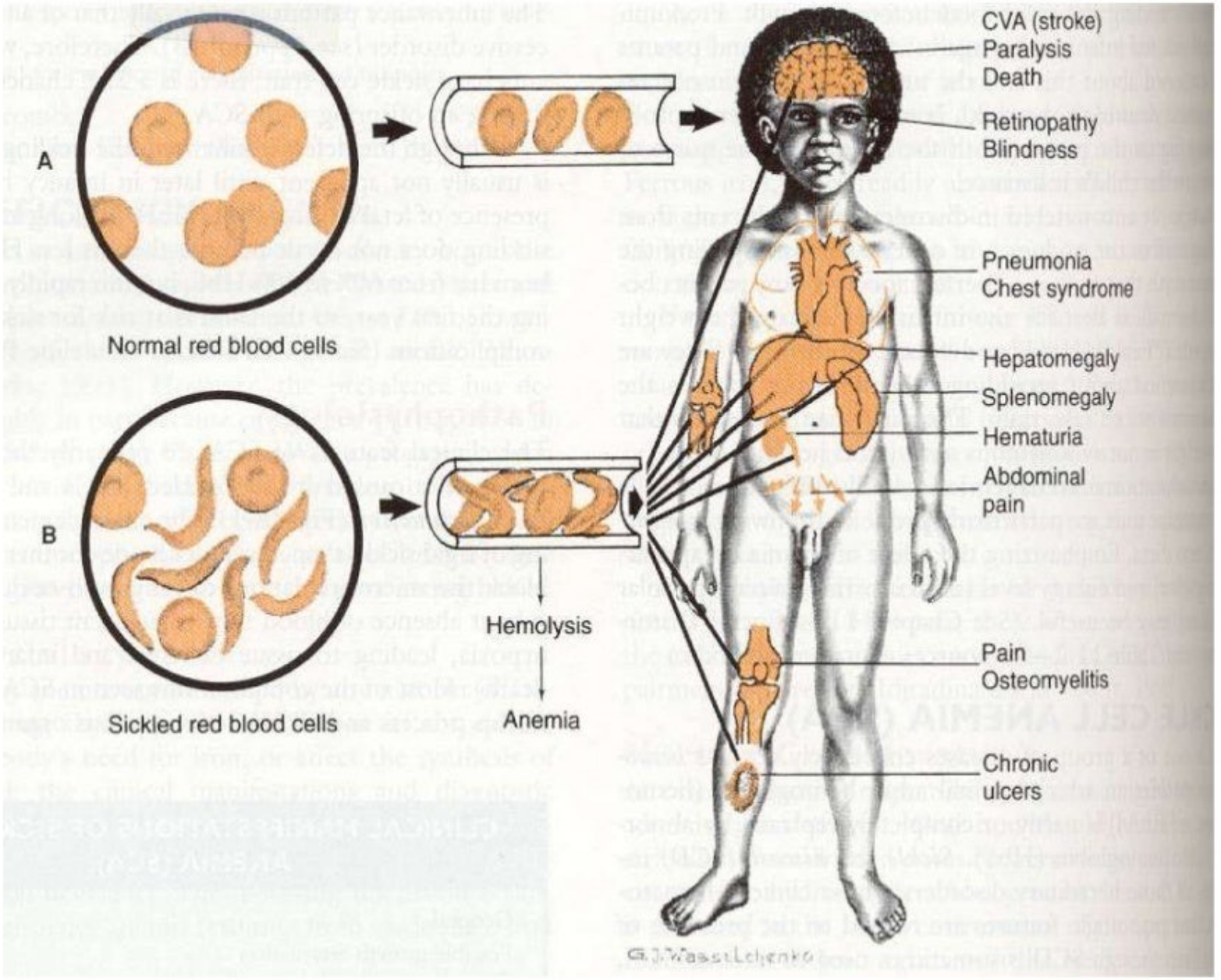
Sickle Cell Trait vs. Disease

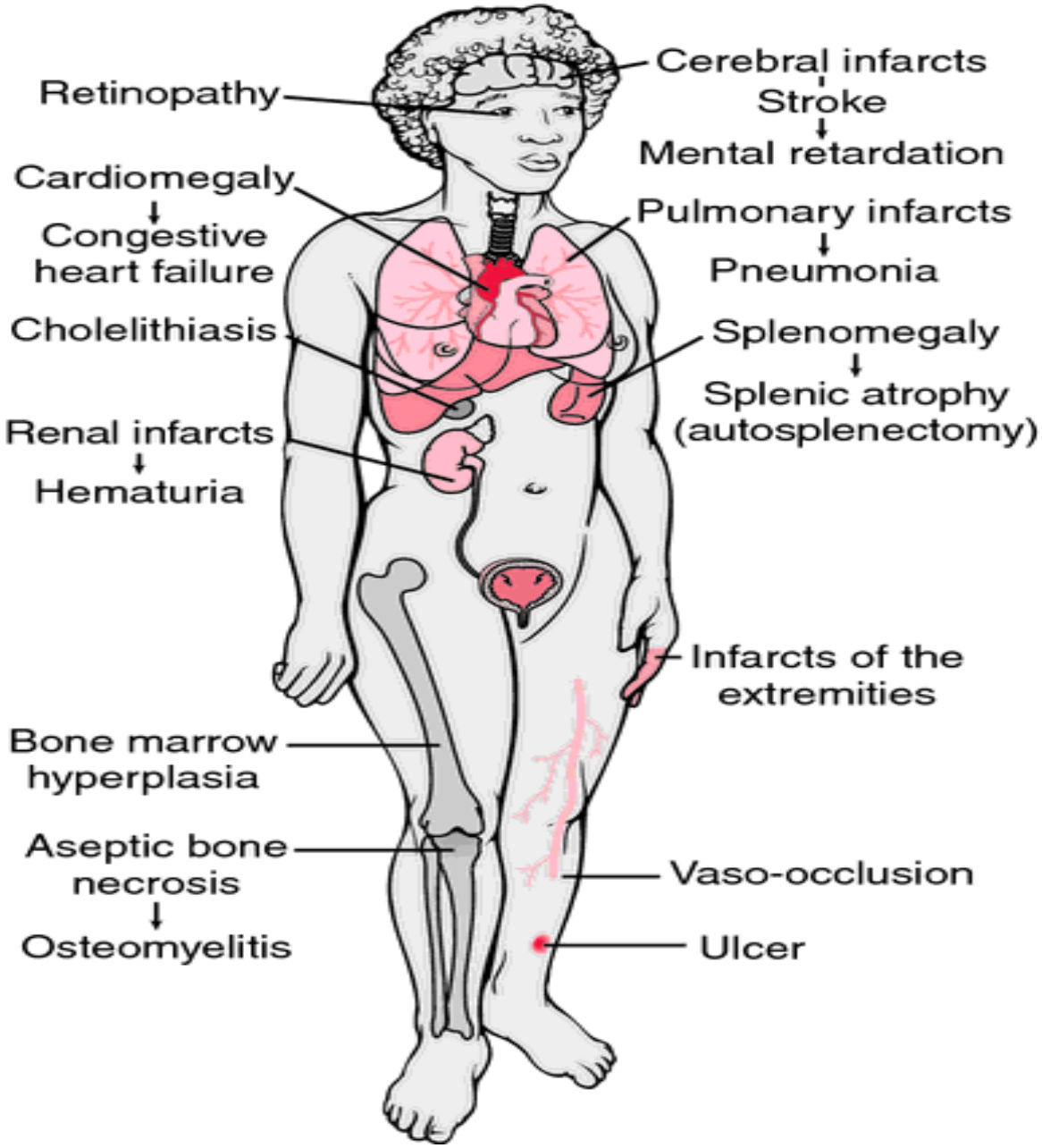
Trait:
Typically no health consequences or symptoms

Disease:
Significantly impacts a person's health and causes many symptoms

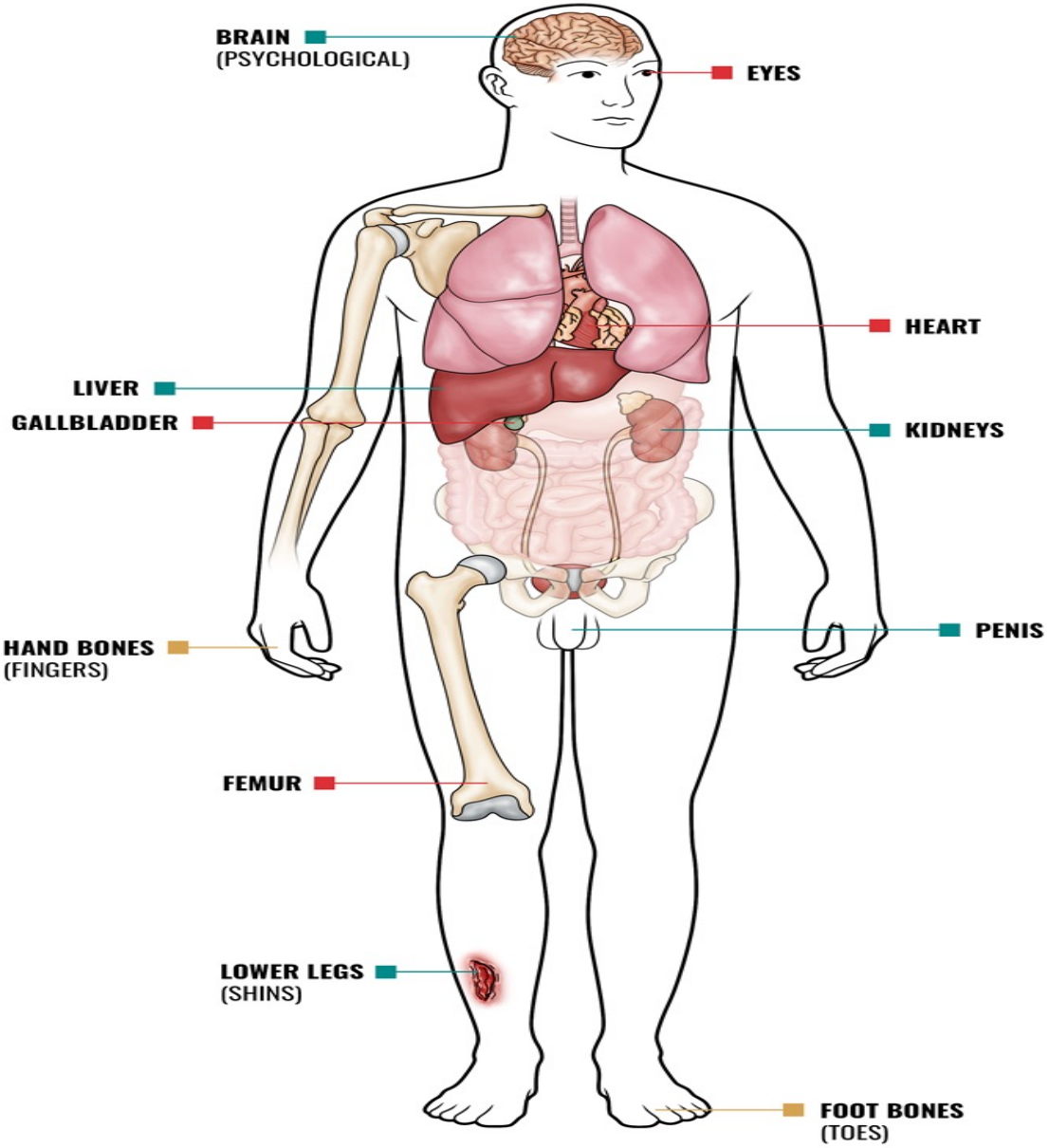





A
Normal red blood cells
B
Sickled red blood cells
Hemolysis
Anemia
CVA (stroke)
Paralysis
Death
Retinopathy
Blindness
Pneumonia
Chest syndrome
Hepatomegaly
Splenomegaly
Hematuria
Abdominal
pain
Pain
Osteomyelitis
Chronic
ulcers



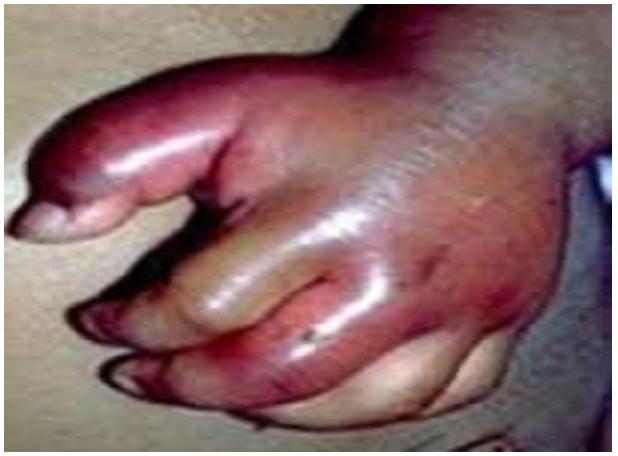
Clinicopathologic findings in sickle cell anemia. The findings are a consequence of in farctions, anemia, hemolysis, and recurrent infection. From Damjanov, 2000.


BRAIN
(PSYCHOLOGICAL)
EYES
HEART
LIVER
GALLBLADDER
KIDNEYS
HAND BONES
(FINGERS)
PENIS
FEMUR
LOWER LEGS
(SHINS)
FOOT BONES
(TOES)
Acute complications
Chronic complications
Both acute and chronic complications

Clinical manifestation

General

- Possible growth retardation
- Chronic anemia (hemoglobin level of 6–9 g/dl)
- Possible delayed sexual maturation
- Marked susceptibility to sepsis



- Vasoocclusive Crisis

Pain in area(s) of involvement

- Manifestations related to ischemia of involved areas
- Extremities—Painful swelling, painful joints
- Abdomen—Severe pain
- Chest—Symptoms resembling pneumonia
- Liver—Obstructive jaundice, hepatic coma
- Kidney—Hematuria
- Genitalia

- **Sequestration Crisis**
- Pooling of large amounts of blood
- Hepatomegaly
- Splenomegaly
- Circulatory collapse
- Heart—Cardiomegaly, systolic murmurs
- Lungs—Altered pulmonary function

Therapeutic Management

- The aims of therapy are to :
- prevent the sickling phenomena, which are responsible for the pathologic sequelae,
- treat the medical emergencies of sickle cell crisis.

Medical management

- of a crisis is usually directed toward supportive and symptomatic treatment.
- **The main objectives are to provide:**
- (1) rest to minimize energy expenditure and to improve oxygen utilization;
- (2) hydration through oral and IV therapy;
- (3) electrolyte replacement because hypoxia results in metabolic acidosis, which also promotes sickling;

- (4) analgesia for the severe pain from vasoocclusion;
- (5) blood replacement to treat anemia
- (6) antibiotics to treat any existing infection.
- (7) Administration of pneumococcal and meningococcal vaccines.
- 8) Oral penicillin prophylaxis is also recommended by 2 months.

- 9) Oxygen therapy is of little therapeutic value unless the patient has hypoxia.
- 10) blood transfusions. Exchange RBC transfusion.
- 11) splenectomy may be a lifesaving measure.

Prognosis

- The greatest risk is usually in children younger than 5 years of age, and the majority of deaths in these children are caused by overwhelming infection.

Nursing Care Management

I. Educate the Family and Child

(1) seek early intervention for problems, such as fever of 38.5° C or greater;

(2) give penicillin as ordered;

(3) respiratory problems that can lead to hypoxia;

(4) The nurse tells the family that the child is normal but can get sick in ways that other children cannot.

(5) the importance of adequate hydration

(6) impaired kidney function result in the problem of enuresis.

Nursing Care Management

II. Promote Supportive Therapies During Crises

1) Management of pain is an especially.
2) Any pain program should be combined with psychologic support to help the child deal with the depression, anxiety, and fear that may accompany the disease.
3) If blood transfusions or exchange transfusions are given, the nurse has the responsibility of observing for signs of transfusion reaction .
4) Record intake, especially of IV fluids, and output. The child's weight should be taken on admission to serve as a baseline for evaluating hydration. Because diuresis can result in electrolyte loss, the nurse also observes for signs of hypokalemia and should be familiar with normal serum electrolyte values to report changes.

Nursing Care Management

II. Promote Supportive Therapies During Crises

5) The nurse should be aware of spleen size because increasing splenomegaly is an ominous sign. A decreasing spleen size denotes response to therapy.
6) Management of pain is an especially.
7) Any pain program should be combined with psychologic support to help the child deal with the depression, anxiety, and fear that may accompany the disease.
8) If blood transfusions or exchange transfusions are given, the nurse has the responsibility of observing for signs of transfusion reaction .
9) Because hypervolemia from too-rapid transfusion can increase the workload of the heart, the nurse also is alert to signs of cardiac failure.
10) Vital signs and blood pressure are also closely monitored for impending shock.

Nursing Care Management

III. Recognize Other Complications

Nurses also need to be aware of the signs of ACS and CVA, both poten- tially fatal complications.

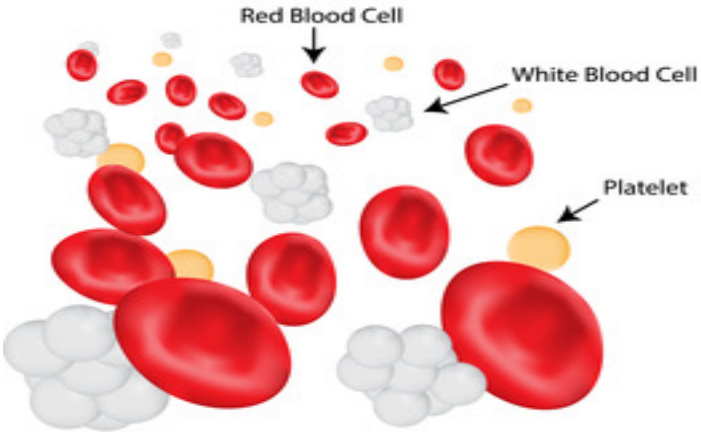
IV. Support the Family

1) Families need the opportunity to discuss their feelings regarding transmitting a potentially fatal, chronic illness to their child. parents express their prevalent fear of the child's death.
2) The nurse should care for the family as for any family with a child who has a chronic and life-threatening illness and give consideration to the siblings' reactions, the stress on the marital relationship, and the childrearing attitudes displayed toward the child .

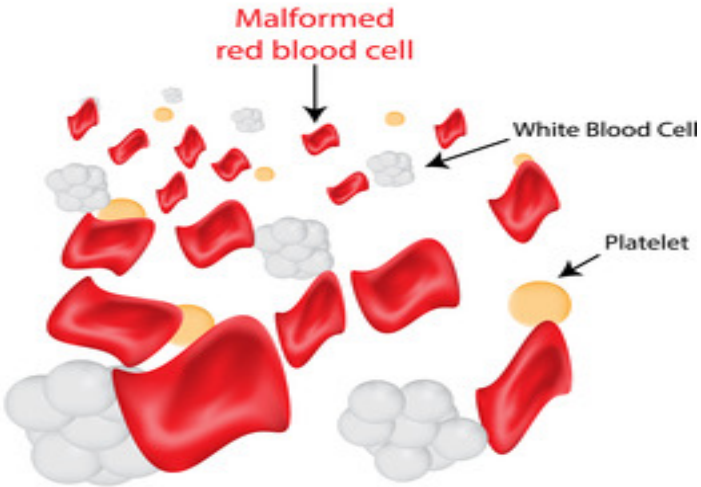
B-thalassemia (COOLEY ANEMIA)

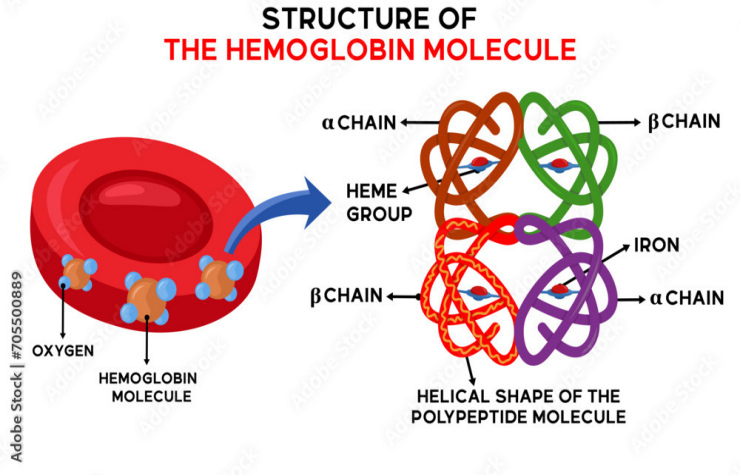
Thalassemia

Normal



Thalassemia





β-THALASSEMIA (COOLEY ANEMIA)

- Worldwide, thalassemia is a common genetic disorder, affecting as many as 15 million people.
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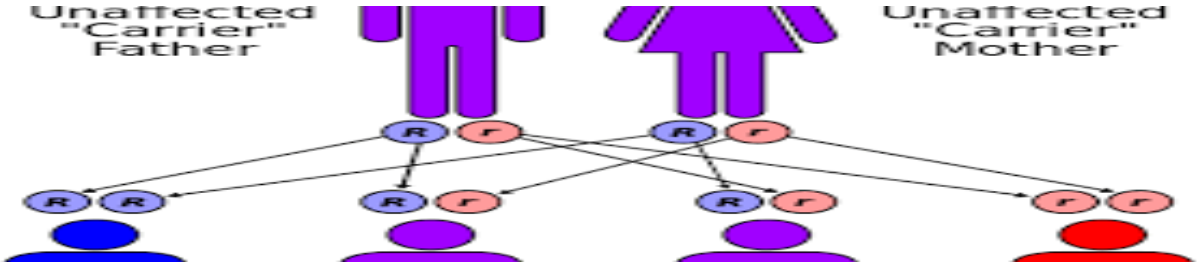
- The term thalassemia, which is derived from the Greek word thalassa, meaning “sea,” is applied to a variety of inherited blood disorders characterized by deficiencies in the rate of production of specific globin chains in Hgb.



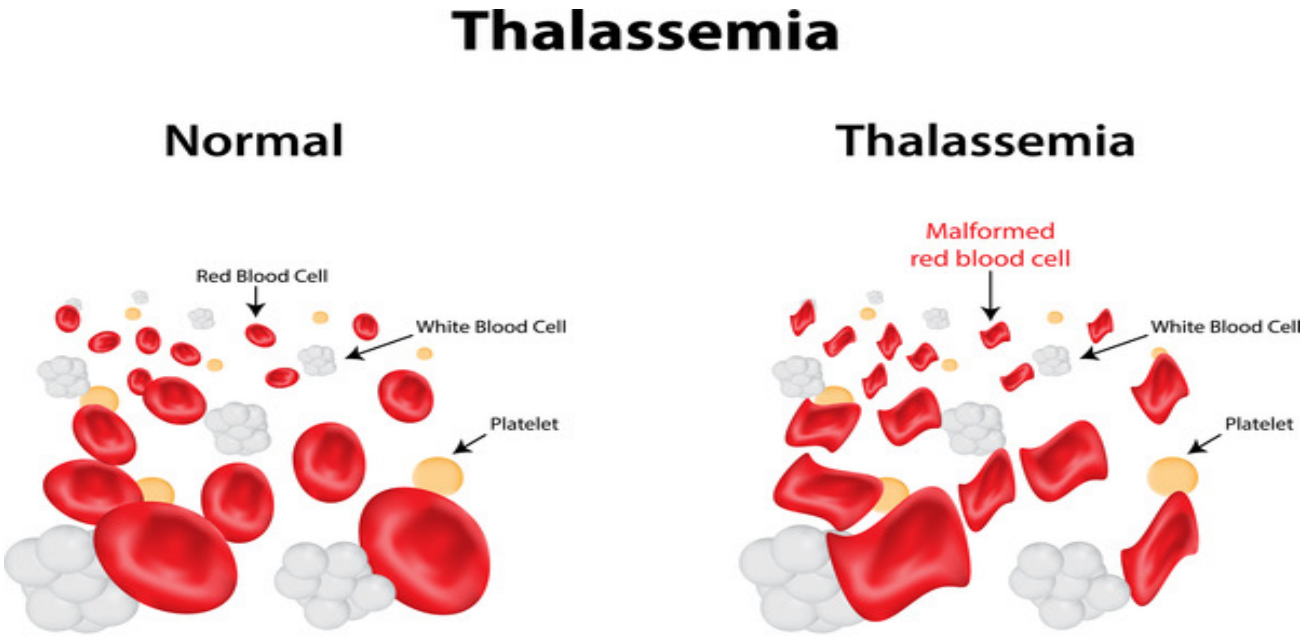
β-Thalassemia is the most common of the thalassemias and occurs in four forms:

Three heterozygous forms:

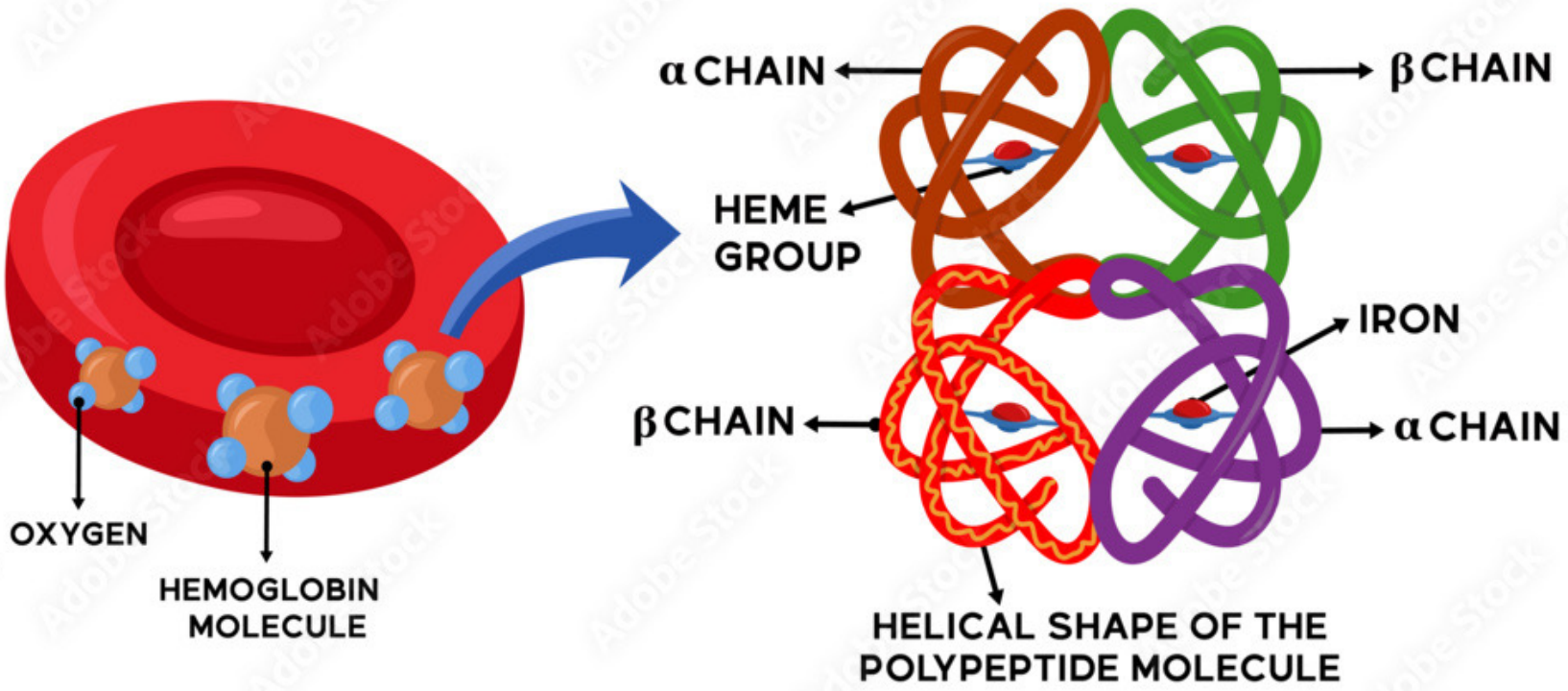
1. **Thalassemia minor**,
2. **Thalassemia intermedia,** which is manifested as splenomegaly and moderate to severe anemia
3. A homozygous form, **thalassemia major** (also known as Cooley anemia),

Pathophysiology

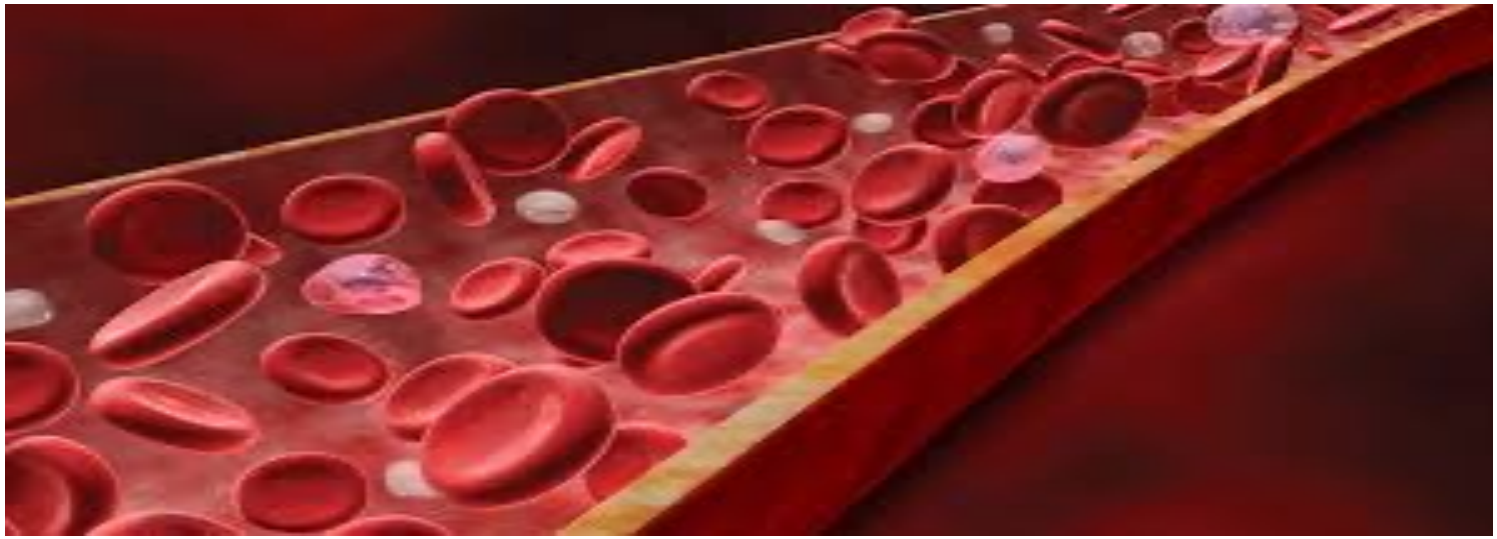
- Normal postnatal Hgb is composed of two α- and two β-polypeptide chains.
- In β-thalassemia, there is a partial or complete deficiency in the synthesis of the **β-chain** of the Hgb molecule.
- Consequently, there is a compensatory increase in the synthesis of α-chains, and γ-chain production remains activated, resulting in defective Hgb formation.



STRUCTURE OF
THE HEMOGLOBIN MOLECULE

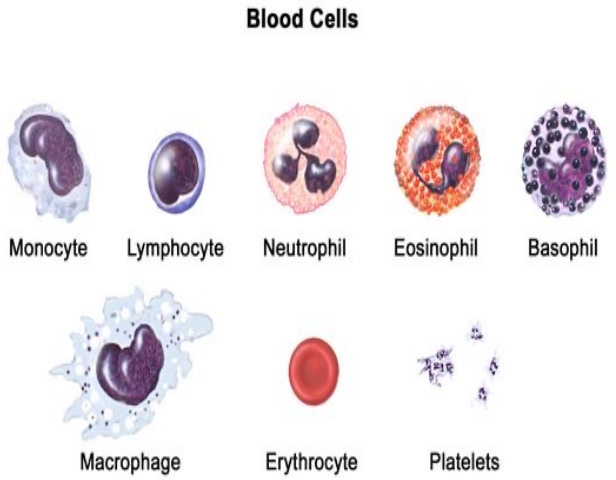


- Excess iron from hemolysis of supplemental RBCs in transfusions and from the rapid destruction of defective cells is stored in various organs (hemosiderosis).



Diagnostic Evaluation

- The onset of thalassemia major may be and not recognized until the latter half of infancy.
- The clinical effects of thalassemia major are primarily attributable to:-
- (1) defective synthesis of HbA,
- (2) structurally impaired RBCs, and
- (3) shortened life span of erythrocytes



CLINICAL MANIFESTATIONS OF β-THALASSEMIA

Anemia (Before Diagnosis)

Pallor
Unexplained fever
Poor feeding
Enlarged spleen or liver

Progressive Anemia

Signs of chronic hypoxia Headache
Precordial and bone pain Decreased exercise tolerance
Listlessness
Anorexia

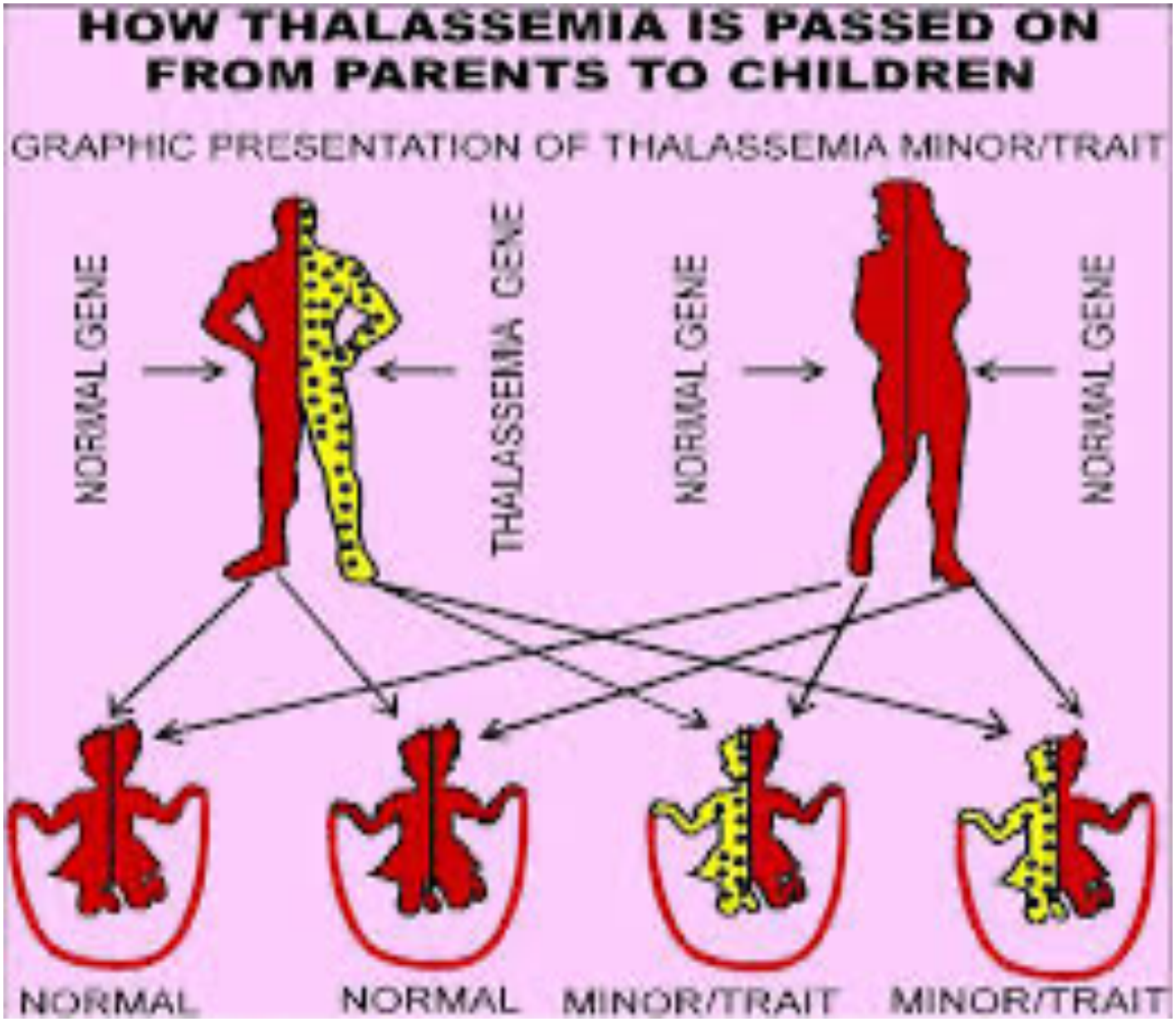
CLINICAL MANIFESTATIONS OF β-THALASSEMIA

Other Features

Small stature
Delayed sexual maturation
Bronzed, freckled complexion (if not receiving chelation therapy)

Bone Changes (Older Children If Untreated)

Enlarged head
Prominent frontal and parietal bossing
Prominent malar eminences
Flat or depressed bridge of the nose
Enlarged maxilla
Protrusion of the lip and upper central incisors and eventual malocclusion Generalized osteoporosis


HOW THALASSEMIA IS PASSED ON FROM PARENTS TO CHILDREN
GRAPHIC PRESENTATION OF THALASSEMIA MINOR/TRAIT
NORMAL GENE
THALASSEMIA GENE
NORMAL GENE
NORMAL GENE
NORMAL
NORMAL
MINOR/TRAIT
MINOR/TRAIT

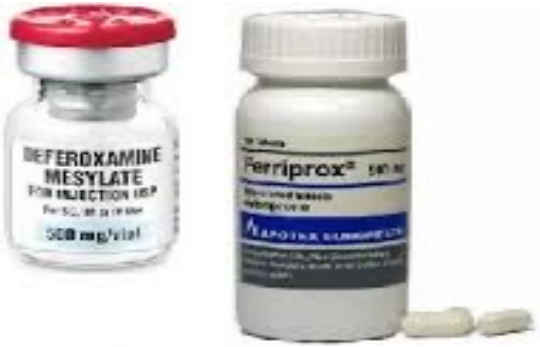
Therapeutic Management

- The objectives of supportive therapy are to maintain sufficient Hgb levels to prevent bone marrow expansion and support normal growth and normal physical activity.
- Transfusions / Hgb level above

9.5 g/dl, an aim that may require

transfusions as often as every

3 to 5 weeks.



The advantages of this therapy include :

(1) improved physical and psychologic well-being

(2) decreased cardiomegaly, hepatosplenomegaly, and fewer bone changes,

(4) normal or near-normal growth and development until puberty,

(5) fewer infections.

6) To minimize the development of hemosiderosis,

- One of the potential complications of frequent blood transfusions is **iron overload (hemosiderosis).**
- Because the body has no effective means of eliminating the excess iron, the mineral is deposited in body tissues.
- To minimize the development of hemosiderosis, the oral iron chelator deferasirox has been shown to be a safe equivalent to **defer-oxamine (Desferal)**, a parenteral iron-chelating agent, and more tolerable by patients and families

7) a splenectomy may be necessary.

After a splenectomy, children generally require fewer transfusions.

A major postsplenectomy complication is severe and overwhelming infection.

8) A curative treatment for some children is Bone marrow transplantation is called hematopoetic stem cell transplantation (HSCT) .

Prognosis

Most children treated with blood transfusion and early chelation therapy survive well into adulthood. The most common causes of death are heart disease, postsplenectomy sepsis, and multiple-organ failure secondary to **hemochromatosis.** A curative treatment for some children is HSCT. Children younger than 16 years of age who undergo allogeneic HSCT have a high rate of complication-free survival; approximately 80% of these children are cured.

Nursing Care Management

The objectives of nursing care are to :

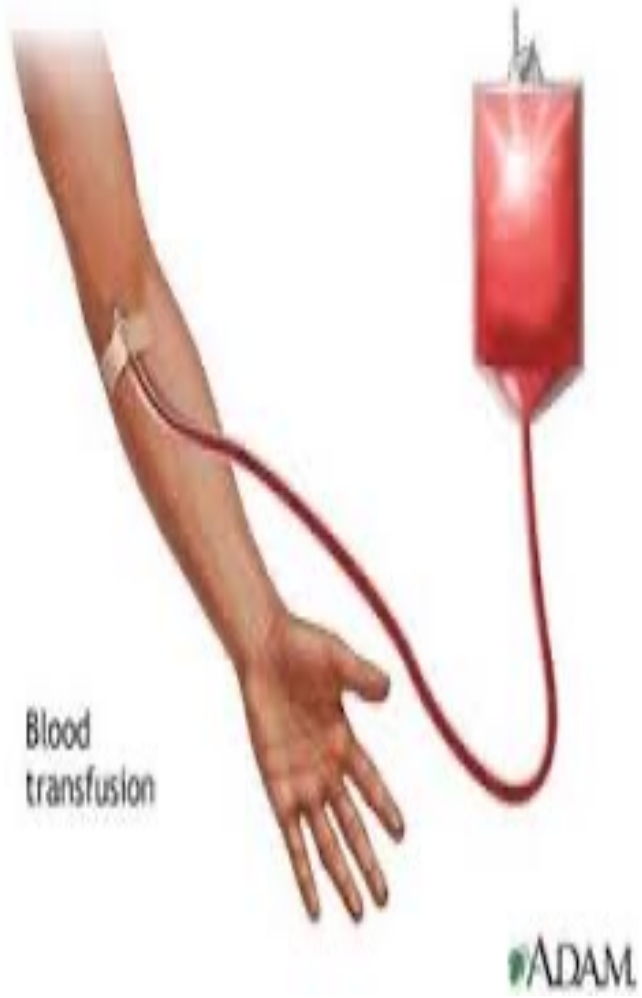
(1) promote compliance with transfusion and chelation therapy.

(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.

5) diet supplementary education.


Blood
transfusion
A.D.A.M.

HEMOPHILIA

HEMOPHILIA

- The term **hemophilia** refers to a group of bleeding disorders in which there is a deficiency of one of the factors (proteins) necessary for coagulation of the blood.
- Although the symptomatology is similar regardless of which clotting factor is deficient, the identification of specific factor deficiencies allows definitive treatment with replacement agents.

- Hemophilia is a rare disorder in which the blood doesn't clot in the typical way because it doesn't have enough blood-clotting proteins (clotting factors).
- Patient with hemophilia, might bleed for a longer time after an injury than normal.

Introduction

- **Hemophilia** is usually an inherited, congenital bleeding disorder characterized by a lack of blood clotting factors, especially factors VIII and IX.

- It is an X-linked disorder primarily affecting males; females act as carriers. Occurs in 1 in 5,000 males.

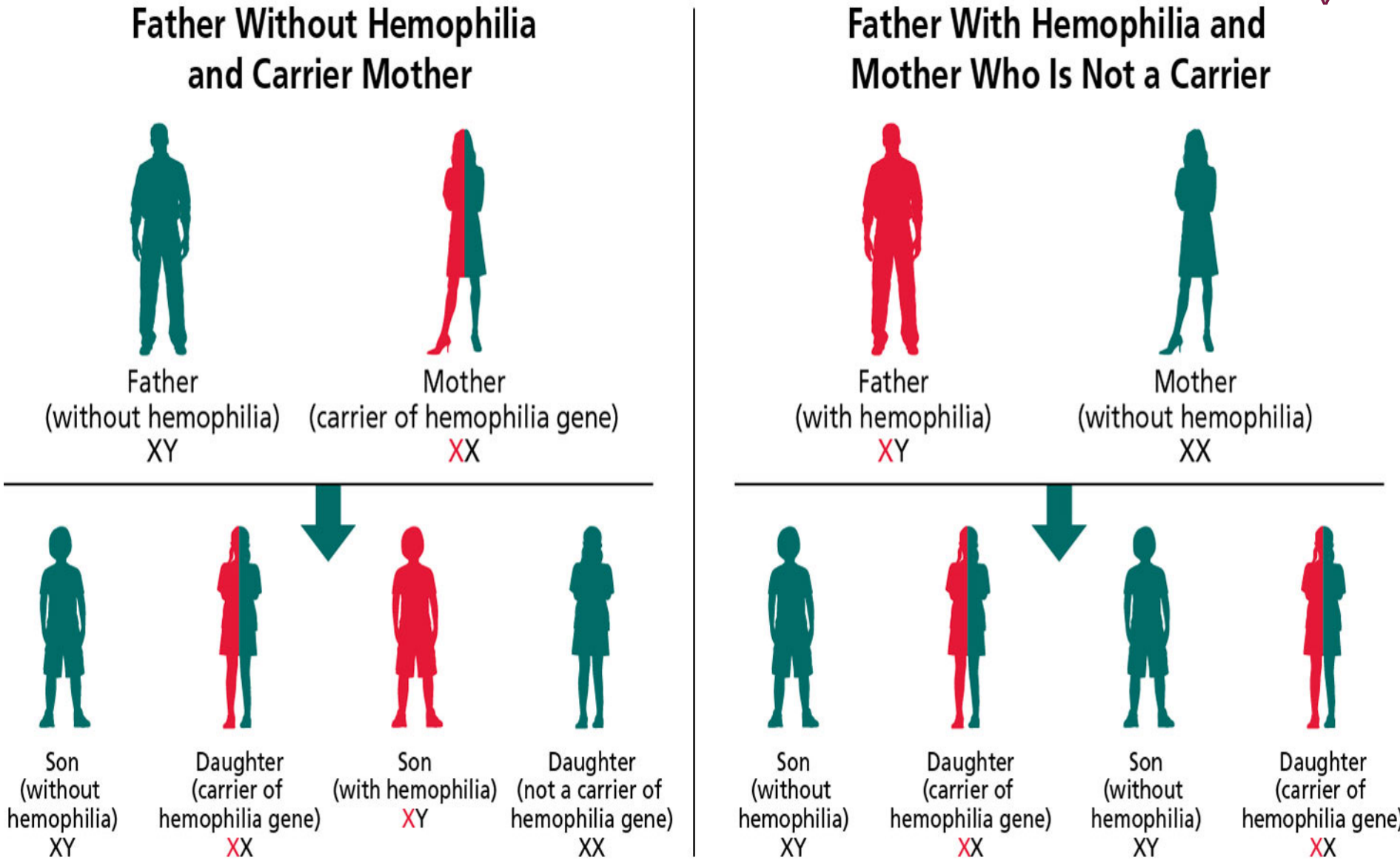
Classification of hemophilia

- 80% to 85% have factor VIII deficiency or hemophilia A (classic hemophilia).
- 15% to 20% have factor IX deficiency or hemophilia B
- Few have factor XI deficiency or hemophilia C.

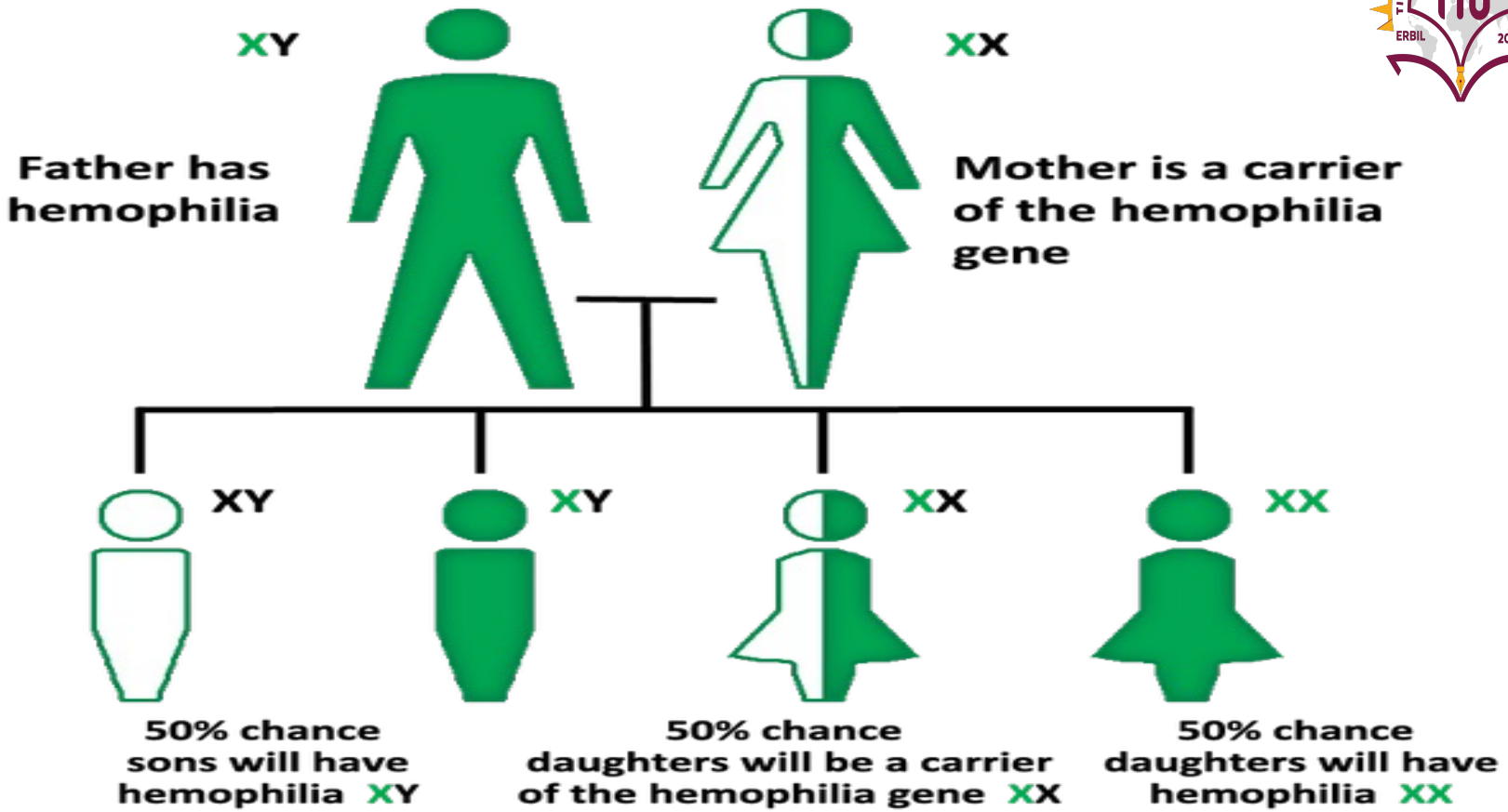
Etiology

- Hereditary (approximately 80% of patients).
- In 1/3 of hemophiliac patients, there is no family history of bleeding.
- The result is an unstable fibrin clot.

Hereditary


Father Without Hemophilia and Carrier Mother
Father (without hemophilia) XY
Mother (carrier of hemophilia gene) XX
Son (without hemophilia) XY
Daughter (carrier of hemophilia gene) XX
Son (with hemophilia) XY
Daughter (not a carrier of hemophilia gene) XX
Father With Hemophilia and Mother Who Is Not a Carrier
Father (with hemophilia) XY
Mother (without hemophilia) XX
Son (without hemophilia) XY
Daughter (carrier of hemophilia gene) XX
Son (without hemophilia) XY
Daughter (carrier of hemophilia gene) XX

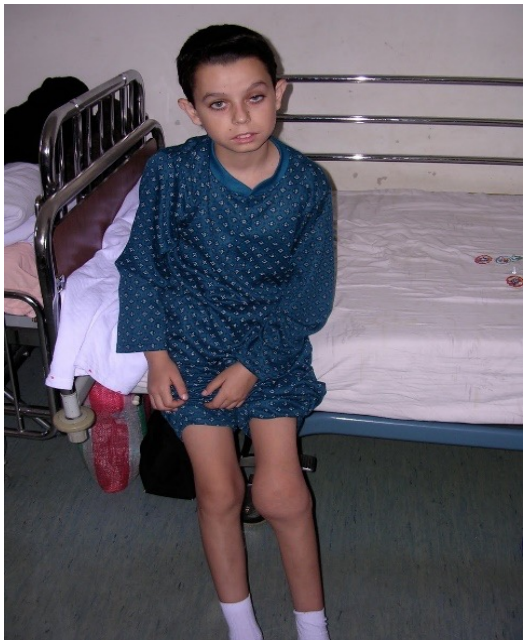
Hereditary



hemophiliafed.org

Pathophysiology and Etiology (Cont.)

- The basic defect is in the intrinsic phase of the coagulation cascade.
- The blood clotting factors are necessary for the formation of prothrombin activator, which acts as a catalyst in the conversion of prothrombin to thrombin.



Injury Occurs

1. Injury to blood vessel results in bleeding.

2. Vessel constricts and clotting factors are activated.

Normal

3. Along with other substances, clotting factor VIII causes a strong platelet plug to form.

4. A stable fibrin clot forms over the platelet plug as a final seal on the injury, and the bleeding stops.

Hemophilia

3. Lack of clotting factor VIII causes a weak platelet plug to form.

4. Incomplete and/or delayed fibrin clot allows bleeding to continue.

How to detect

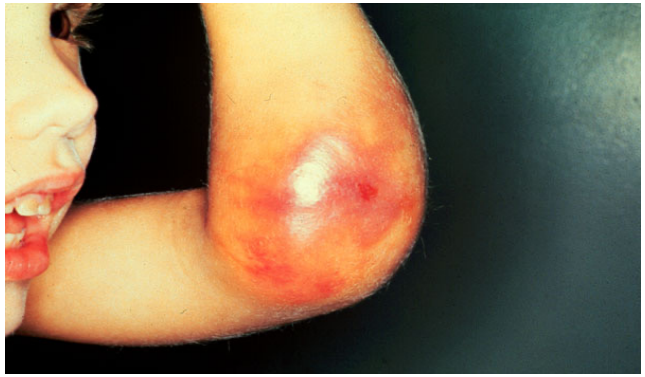
- Seldom diagnosed in infancy unless excessive bleeding is observed from the umbilical cord or after circumcision.
- Usually diagnosed after the child becomes active.

Clinical manifestations :

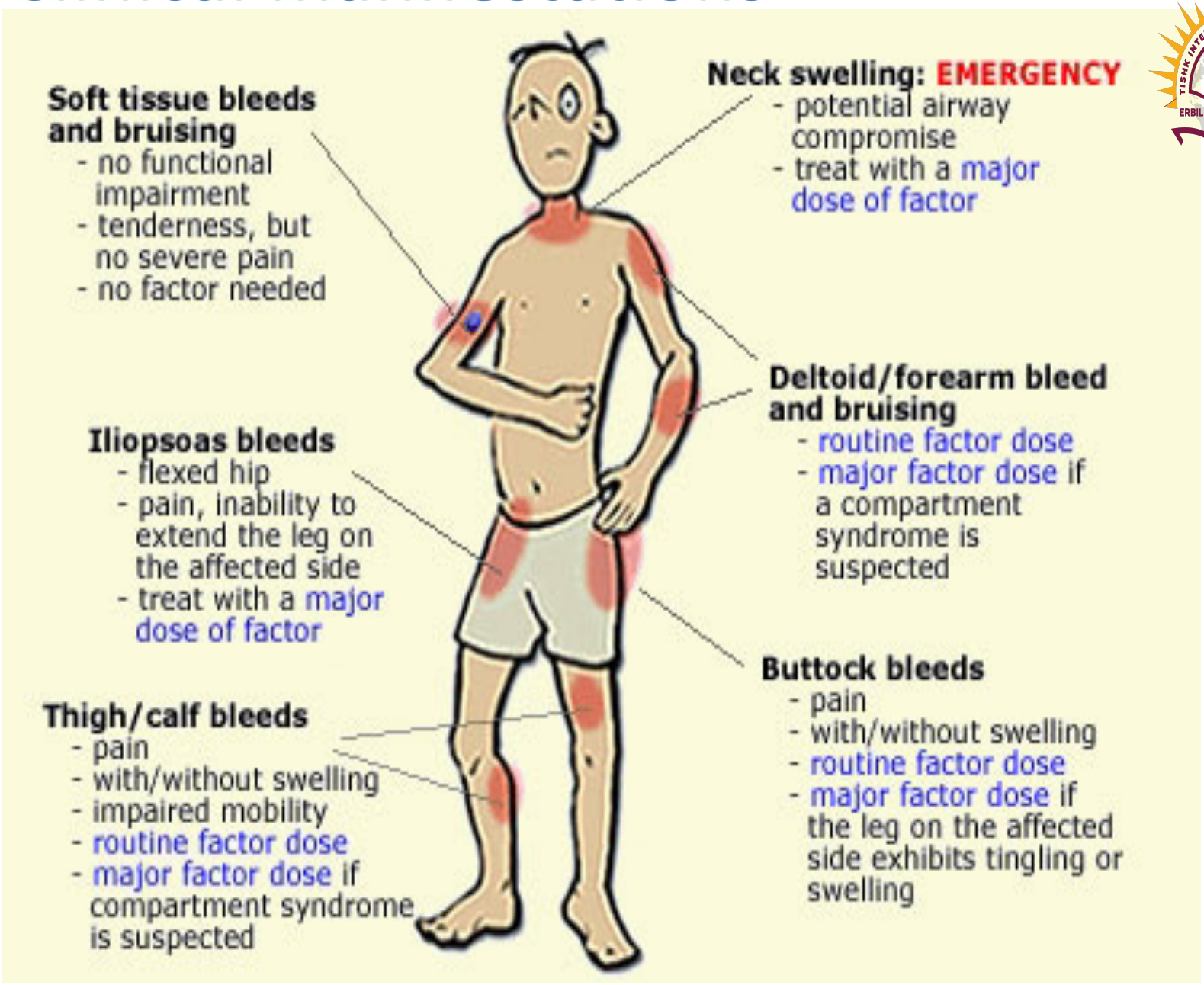
- History of prolonged bleeding occurrences such as after circumcision.
- Easily bruised.
- Prolonged bleeding from the mucous membranes of the nose and mouth from lacerations.
- Hemorrhages into the joints (hemarthrosis) , causing pain, swelling, limitation of movement.

Clinical manifestations

- Spontaneous soft tissue hematomas.
- Cyclic bleeding episodes may occur with periods of little bleeding, followed by periods of severe bleeding.
- Head trauma, resulting in intracranial hemorrhage.
- GI bleeding.



Clinical manifestations



Diagnostic Evaluation

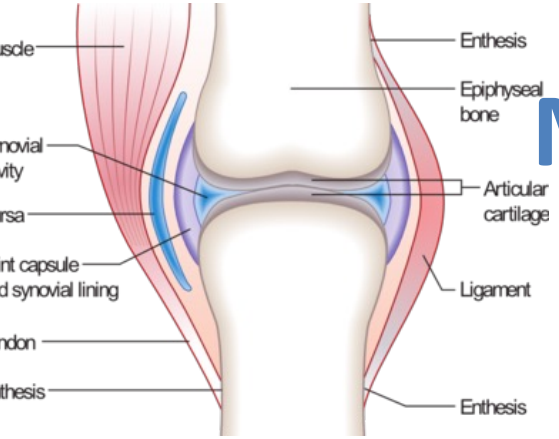
- Test for Prothrombin time and bleeding time.
- Partial thromboplastin time.
- Thromboplastin .
- Assays for specific clotting factors abnormal.
- Gene analysis to detect carrier state, for prenatal diagnosis.

Management

- Must replace coagulation factor (VIII or IX).
- Rapid, early, appropriate treatment is the key to preventing most complications.
- fresh frozen plasma is given to supply factor XI.
- Antifibrinolytics, such as aminocaproic acid and tranexamic acid are given as adjunctive therapy for mucosal bleeding to prevent clot breakdown by salivary proteins.

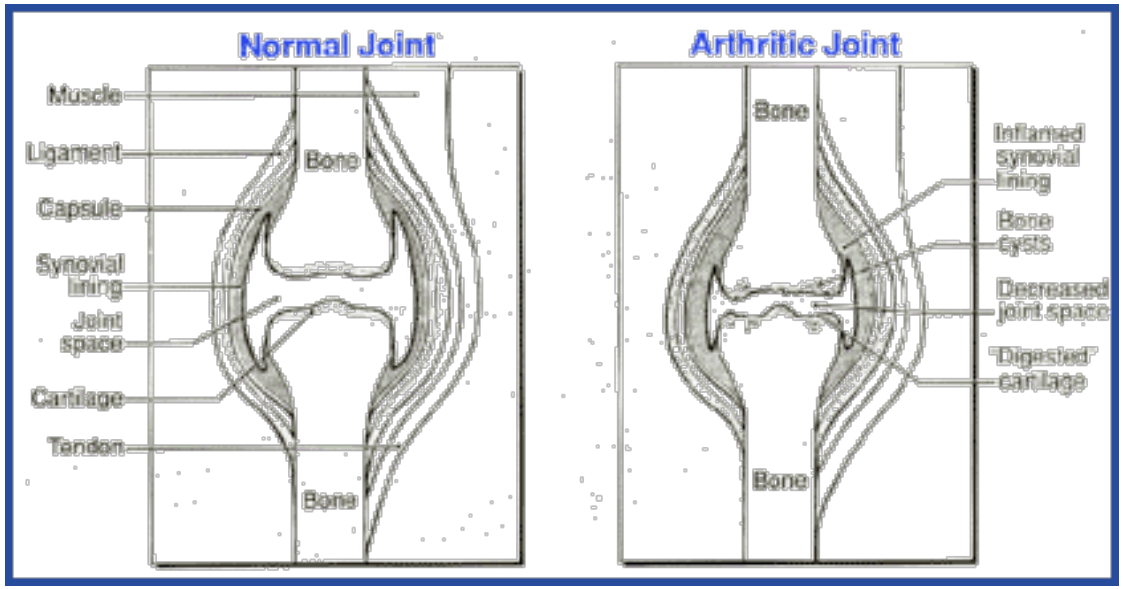
Management

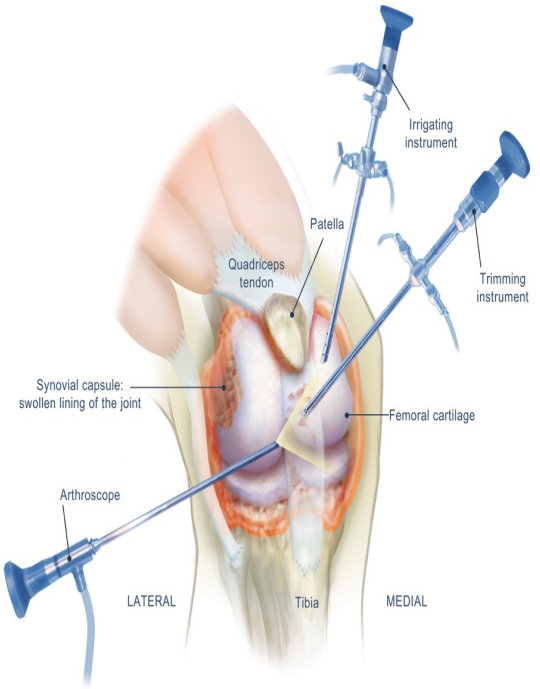
- Supportive therapies:
  - NSAIDs are used to decrease inflammation and arthritic-like pain associated with chronic hemarthroses. Must be used with caution because some types and higher doses interfere with platelet adhesion.
  - Physical therapy to prevent contractures and muscle atrophy. This includes exercise, whirlpool, and icing.
  - Orthotics to prevent injury to affected joint and to help resolve hemorrhages.



Management

- Synovectomy orthopedic surgical intervention to remove damaged synovium in chronically involved joints.





Complications

- Airway obstruction
- Intestinal obstruction
- Compression of nerves with paralysis
- Repeated hemorrhages may produce degenerative joint changes with osteoporosis and muscle atrophy.
- Intracranial bleeding, resulting in serious neurologic impairments.

Complications:

- Hepatitis
- Uncertain life span .
- Contaminated cryoprecipitate, fresh frozen plasma.
- Death may result from exsanguination after any serious hemorrhage, such as intracranial, airway, or other highly vascular areas.

Nursing Interventions

- **Provide emergency care for bleeding.**
  - Apply pressure and cold on the area for 10 minutes to allow clot formation.
  - Immobilize the affected part and elevate above the level of the heart.
  - Administer recombinant factor VIII or factor IX coagulation concentrate.

Provide emergency care for bleeding.

- Place fibrin foam or absorbable gelatin foam in the wound.
- Apply fibrinolytic agents to wound for oral bleeding.
- Keep child quiet during treatment to decrease pulse and rate of bleeding.
- Monitor vital signs and treat for shock if child becomes hypotensive

Nursing Interventions

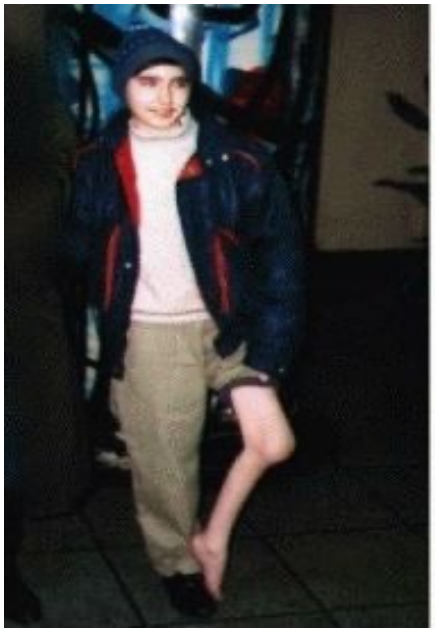
- Providing Protection Against Bleeding
- Avoid rectal temperatures
- **Avoid injections if possible.**

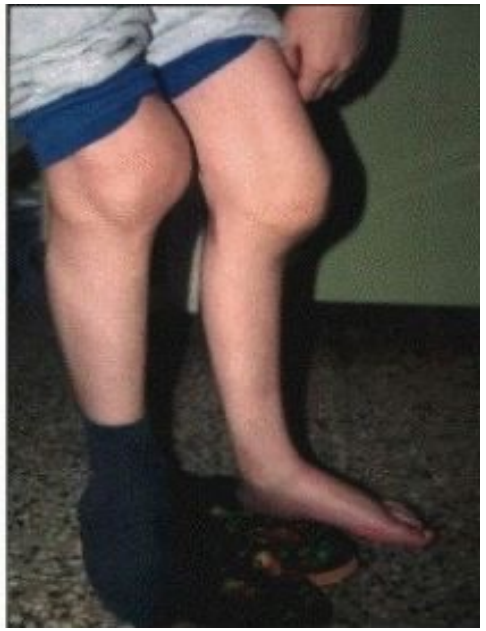
1. Administer medications orally whenever possible.
2. Subcutaneous route is preferred over I.M.
3. Apply pressure to injection site for 10 to 15 minutes. Then apply a pressure dressing with self-adhesive gauze.

Preserving Mobility

- **Provide supportive care for hemarthrosis.**

1. Immobilize the joint in a position of slight flexion.
2. Elevate the affected part above the level of the heart.
3. Apply ice packs.





Preserving Mobility (Cont.)

- For severe hemarthroses, continue immobilization through casting, if necessary, and prevent weight bearing on the affected limb.
- Administer short course of corticosteroids as ordered to relieve inflammation.

Preserving Mobility (Cont.)

- Refer for physical therapy if persistent deformity or specialized treatments are needed.
- For less severe hemarthroses, begin gentle, passive exercise 48 hours after the acute phase to prevent joint stiffness and fibrosis. Progress to active exercises.

Enhancing Family Coping

- Help parents understand that no one is to blame.
- Allow the child and other family members to handle equipment used in care.
- Use play to help the young child and siblings adjust to illness by (transfusing)teddy bear.

Enhancing Family Coping (Cont.)

- Encourage the child's continuing education. Parents fear sending the child to school, but safety issues can be addressed through discussions with the school nurse, teachers, and principal. Home or hospital tutoring should be continued while child is away from school.

Enhancing Family Coping (Cont.)

- Encourage the parents to allow the child to participate in as many normal activities as possible within the realm of safety.
- Refer to social worker for counseling and identification of resources for financial concerns.
- Encourage involvement in support group.

Enhancing Family Coping (Cont.)

- **Encourage avoidance of overprotection**
  - Promote a sense of independence and self-care within the patient's limitations.
  - Encourage healthful activity and reasonably aggressive hobbies. Reinforce self-judgment of child or teenager in selection of safe physical activities.

Encourage avoidance of overprotection (Cont..)

- Participate in as many age-appropriate activities as possible.
- Help parents understand the importance of vocational guidance for their child emphasis should be given to occupations using intellect or skills rather than physical effort.

Community and Home Care Considerations

- **Provide teaching and referrals to initiate an infusion therapy program at home when hemorrhage begins. Assist with teaching the following:**
  - Storage and preparation of replacement factors.
  - Awareness of signs of trans management.
  - Venipuncture technique.
  - Transfusion management.
  - Record keeping.

Community and Home Care Considerations

- **Perform a home safety survey to identify potential hazards to the hemophiliac child, such as**

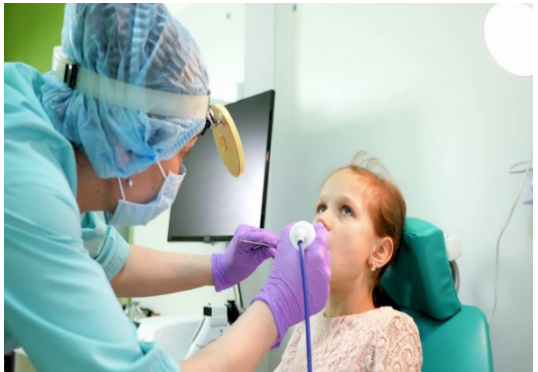
1. cluttered furniture the child may bump into, sharp edges on furniture or other objects.
2. loose rugs that promote falls.
3. slippery tub or floor surfaces.

Community and Home Care Considerations

- Provide education to family and all caregivers about recognizing and treating bleeds appropriately.
- Provide emotional support through the provision of educational materials, information about support groups, and a list of resources within the community.

Educate parent with child to visit the primary health care:

- Regular visits to their primary care provider for preventive health maintenance, to address growth and development, behavior, and psychosocial issues.
- Dental examinations and teeth cleaning every 6 months.
- All the recommended childhood immunizations, plus hepatitis .



Educate teachers and other school faculty about the child's special needs

- These children can have permanent mental or physical disabilities from old hemorrhages, necessitating individualized plans.
- They must avoid rough or contact sports and activities

Educate teachers (Cont.)

- All injuries must be taken seriously. The school nurse should be notified so that proper first aid and treatment can be initiated

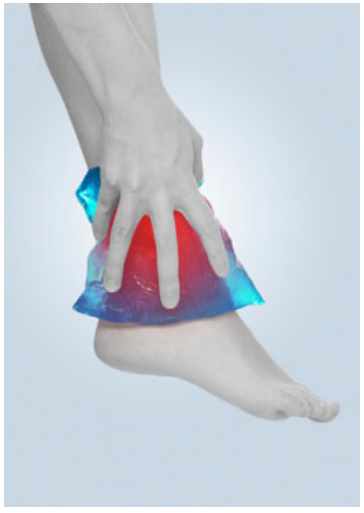
Family Education and Health Maintenance

- Review safety measures to prevent or minimize trauma.

- Remind parents not to administer **aspirin** to the child.

Family Education (Cont.)

- **Teach emergency treatment for hemorrhage.**
  - Immobilize the part with splints or an elastic compression bandage.
  - Apply ice packs. Parents should keep two or three plastic bags of ice immediately available in the freezer.
  - Consult the child's health care provider and initiate additional recommended therapy.



Family Education (Cont.)

- **Encourage regular medical and dental supervision.**
  - Preventive dental care is important. sponge-tipped toothbrushes should be used to prevent bleeding.
  - Hepatitis B vaccine is necessary to protect against hepatitis from blood transfusions.

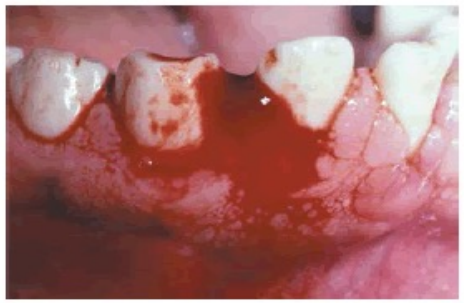


Figure 1. Intraoral bleeding in a hemophilic patient.

Family Education (Cont.)

- Teach healthy diet to avoid obesity, which places additional strain on the child's weight-bearing joints and predisposes to hem-arthroses.
- educate the child to avoid sharp instruments, hard candy, suckers, and other foods with sharp edges that may cause mucosal lacerations.

Family Education (Cont.)

- Assist parents in teaching the child to understand the exact nature of the illness as early as possible. Special attention should be given to the signs of hemorrhage, and the child should be told of the need to report even the slightest bleeding to an adult immediately.
- Educate parents for family planning.

Reference

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2. Hemophilia | National Heart, Lung, and Blood Institute (NHLBI) [Internet]. [cited 2018 Apr 3]. Available from: https://www.nhlbi.nih.gov/health-topics/hemophilia

Thank you