Pediatric Hematologic Disorders NCLEX Review

- Blood cells are produced in the bone marrow beginning with the stem cell, which then become a lymphoid cell that then becomes a lymphocyte (B or T) myeloid cell, which changes to either an erythrocyte (red blood cell) or a granulocyte or monocyte (white blood cells) or megalokaryocyte (platelets precursor). Intrinsic factor stimulates the formation of blood cells (hemopoiesis) cell growth.
- The spleen, which is located in the left upper quadrant of the abdomen, is responsible for filtration of the blood, break down of hemoglobin, removal of old white blood cells, and storage of red blood cells and platelets.
- The liver is the primary site for the production of clotting factors. The liver uses vitamin K to produce prothrombin and factors VII, IX, and X.
- Normal coagulation/clotting involves a local response of vasoconstriction and release of a factor to stimulate platelet adhesion (sticking together) to form a plug and stop bleeding. Clotting factors act to stimulate formation of a fibrin clot by way of an

1. Intrinsic pathway with factor XII, factor XI, I, II, V, VIII, IX, X, high molecular weight kininogen (HMK), and prekallikrein (KAL). Partial thromboplastin time (PTT) is used to measure function of factors.
2. Extrinsic pathway with factor VII, I, II, V, VII, and X. Prothrombin time (PT) measures the function of factors in this pathway.

ANEMIA

A low red blood cell count, including a low hematocrit or hemoglobin level, results in a state referred to as anemia.With decreased blood cells the delivery of oxygen and nutrients is decreased, resulting in poorly nourished or poorly oxygenated body cells and malfunction of body organs and systems. Anemia results when the bone marrow that produces blood, or the kidney that stimulates blood production, is damaged or suppressed. Anemia can also result when a nutritional component needed to form blood, iron, is insufficient, or when blood cells that are produced are poorly structured, sickled, and malfunction.

II. APLASTIC or HYPOPLASTIC ANEMIA

The bone marrow can be damaged due to exposure to radiation, infections (human parvovirus, hepatitis), toxic substances, including radiation or medications administered to suppress cancer cells or eradicate microorganisms, or can result from unknown causes. The condition can be primary (congenital) or secondary (acquired) possibly due to autoimmune disease. The damage to the bone marrow results in the decreased production of white blood cells, red blood cells, and platelets. Hypoplastic anemia results in low red blood cells with either normal or decreased white blood cell count or platelets. Decreased white blood cells (leukocytopenia) place the client at risk for infection, low red blood cells results in decreased cell oxygenation and nutrition, and low platelets can predispose a client to bleeding. Prognosis depends on the extent and duration of decreased blood cell production and client vulnerability due to chronic disease or debility.

Assessment:

- Weakness and fatigue due to deoxygenated, malnourished body tissues
- Pallor due to decreased red blood cells
- Infections due to low white blood cells
- Bruising (ecchymosis) secondary to low platelet count (thrombocytopenia)
- Hemorrhage: Small superficial bleed (petechiae); Nosebleed (epistaxis); Other mucosal bleeding (oral, gastrointestinal, vaginal).

Diagnostics:

- Bone marrow aspiration may reveal fatty yellow bone marrow.
- Complete blood count values reveal low red blood cell count, low white blood cell count, low hemoglobin, low hematocrit, and low platelet counts.
- Pulse oximetry and blood gases may reveal hypoxia in severe anemia.
- Acidosis may result in a decreased serum pH level.
- Electrolyte imbalance may be noted due to acidosis

Treatment:

- Bone marrow replacement/transplant to replace stem cells
- Immunosuppressive therapy to suppress autoimmune response:

1. Antilymphocyte globulin (ALG) Antithymocyte globulin (ATG)
2. Cyclosporine A(CSA)
3. Granulocyte macrophage colony-stimulating factor (GM-CSF)
4. Cyclophosphamide for immunosuppression

- Androgens may be added to ATG to stimulate erythropoiesis.
- Red blood cell transfusion with severe anemia.
- Platelet transfusion if decreased platelet level is severe

NCLEX!!! Chemotherapeutic drugs can result in nausea and vomiting, alopecia, and mucosal ulceration and thus support measures should be taken.

Nursing Interventions:
• Reinforce physician’s explanation of diagnosis and treatment plan.

- Explain procedure at child’s level of understanding including what will be seen, felt, heard, and smelled; use drawings when appropriate.
- Provide antiemetic and appetite stimulant to increase nutritional intake.
- Offer foods after antiemetic takes effect to reduce nausea and maximize caloric intake.
- Allow to eat any food that is tolerated; avoid forcing food during nausea episode. • Rinse mouth to remove unpleasant taste sensation.
- Maintain contact after discharge and between remissions to encourage follow-up care and respond to questions or provide emotional support.
- During intravenous administration of ATG, monitor site closely to prevent infiltration or extravasation.
- Maintain careful asepsis to prevent infection.

- Meticulous mouth care with soft toothbrush to prevent infection, irritation, and bleeding from oral ulceration.
- Liquid, bland, or soft diet as tolerated.

III. IRON DEFICIENCY ANEMIA

Inadequate intake or excessive loss of iron causes this widespread nutritional disorder. Causes of this disorder could include:

- Decreased supply due to poor eating habits, excessive milk or extended breastfeeding, and delayed solid food intake or rapid growth rate
- Inadequate stores of iron at birth, found in low birthweight babies, maternal iron deficiency, or fetal blood loss
- Impaired absorption due to presence of inhibitors such as gastric alkalinity or malabsorption disorders such as lactose intolerance or inflammatory bowel disease or chronic diarrhea
- Increased body need for iron due to prematurity, adolescence, or pregnancy
- Loss of iron due to parasites or blood loss One molecule of the heme in hemoglobin contains an atom of iron; thus insufficient iron results in deficient hemoglobin production.

Assessment:

- Infant may appear underweight due to malnourishment or overweight due to intake of excessive milk with minimal solid food ingestion.
- Characteristic symptoms: irritability, glossitis, stomatitis, koilonychias (concave/spoon fingernails).
- Plasma protein leakage noted with edema, growth retardation.
- Poor muscle development.
- Weakness and fatigue due to deoxygenated, malnourished body tissues.
- Pallor due to decreased red blood cells.
- Infections due to low white blood cells.
- Hemorrhage due to decreased platelets: Small superficial bleed (petechiae); Nosebleed (epistaxis); Bruising (ecchymosis); Other mucosal bleeding (oral, gastrointestinal, vaginal)
- Tachycardia and tachypnea may be present due low blood levels and need to circulate blood more frequently to oxygenate body cells.

Diagnostics:

- Complete blood count values reveal low red blood cell count, low white blood cell count, low hemoglobin, low hematocrit, and low platelet counts.
- Decreased serum proteins, albumen, transferrin, and gamma globulin.
- Reticulocyte count normal or slightly reduced.
- Serum iron concentration (SIC) about 70 mg/dL; total iron-binding capacity to detect transferrin iron binding globulin (TIBG) usually elevated >350 mg/dLfor children 6 months to 2 years of age or >450 mg/dL for persons >2 years of age.
- Transferrin saturation—SIC divided by TIBC multiplied by 100—if below 10% = anemia.
- Guaiac stool to detect chronic bleeding.
- Pulse oximetry and blood gases may reveal hypoxia in severe anemia.

- Acidosis may result in a decreased serum pH level.
- Electrolyte imbalance may be noted due to acidosis.

Treatment:

Prevention with nutrition:

- Breast milk or iron-fortified milk during first year.
- Iron supplement with milk or iron-fortified cereal by age 4 to 6 months (2 months of age in premature infants).
- Iron drops to breast-fed premature infants after 2 months of age.
- Limit formula to 1 L/day and encourage iron-rich solid foods.
- Avoid fresh cow's milk to avoid allergy and gastrointestinal blood loss.
- Supplemental iron (intramuscular [IM] or intravenous [IV] if unable to absorb gastrointestinally).
- Iron-fortified cereal.
- Vitamin B12 IM to treat deficiency due to failure of gastric mucosa to secrete intrinsic factor needed to absorb vitamin B12 (pernicious anemia more common in adults).
- Packed red blood cells if anemia severe.
- Oxygen supplement if severe hypoxia noted.

Nursing Interventions:

- Monitor vital signs for signs of circulatory or respiratory distress due to low blood levels and poor oxygenation.
- If tolerated, administer oral iron compound between meals because high stomach acid enhances absorption:

1. Ferrous sulfate
2. Ferrous gluconate
3. Ferrous fumarate

- Administer iron with meals to reduce nausea and diarrhea (if necessary).
- Parenteral (IM or IV) iron if unable to absorb oral dose: Iron dextran IM or IV, Iron sodium gluconate IV, Iron sucrose complex IV
- Request stool softener as indicated to treat constipation from iron.

NCLEX!!! Liquid iron agents should be taken with a straw to avoid contact with teeth and resulting staining.

Client and Family Teaching:

- Teach proper nutrition as per treatment plan.
- Inform family of dietary sources high in iron such as green leafy vegetables.
- Administer with juices because vitamin C enhance absorption.
- Stress the importance of follow-up blood testing to determine if hemoglobin and hematocrit are adequate and iron administration is effective.
- Teach family to administer iron properly:

1. Oral medication with straw if liquid form is administered; avoid substances that impair absorption (tea, antacid, milk).
2. Teach the Z-track method for IM injections.

- Caution family and client that stool will be dark green to black due to iron content.

NCLEX!!! Use Z-track injection to prevent staining of the skin.

NCLEX!!! Avoid administering iron with tea, antacid, or milk to maximize absorption.

IV. SICKLE CELL ANEMIA

In sickle cell anemia (SCA)/Hgb SS cell disease, an abnormal gene results in production of an irregular red blood cell called hemoglobin (Hgb) S that replaces some of the normal hemoglobin A. The red blood cells collapse into a crescent shape (sickling) when stressed such as during dehydration, hypoxemia, or acidosis. When cells sickle, clumping is noted that obstructs small blood vessels and blocks blood flow. These cells also have a short lifespan, resulting in early destruction due to damaged cell membrane and low blood count: anemia. This condition is an autosomal recessive condition requiring the gene from both parents. Some clients inherit one gene and may exhibit the sickle cell trait, which may or may not be symptomatic under severe conditions such as hypoxia during exertion in low-oxygen settings (high altitude). Clients of African descent have a high incidence of sickle cell anemia. Sickle cell anemia is a chronic illness with distress resulting from blocked and inadequate circulation and tissue/organ damage that cause pain and over time organ failure and death.

Assessment:

- Acute pain due to blocked blood vessels and tissue ischemia, found in:

1. Extremities: swelling of hands, feet, and joints—dactylitis (handfoot syndrome)
2. Chest: pain and pulmonary disease
3. Liver: jaundice and hepatic coma
4. Kidney: hematuria and impaired function
5. Brain: stroke
6. Genitalia: painful erection (priapism)

- Crisis episodes due to:

1. Vasoocclusion: most common crisis due to blocked blood flow from sickling
2. Sequestration
3. Aplastic crisis due to extreme drop in red blood cells (RBC) (often viral trigger)
4. Megaloblastic anemia with excess need for folic acid or vitamin B12 resulting in deficiency
5. Hyperhemolytic crisis—rapid RBC destruction—anemia, jaundice, and reticulocytosis

- Sickling episodes have exacerbation with remissions after effective treatment.
- Fatigue secondary to the anemia.
- Fever during a sickling episode possibly due to infection that provoked distress.

- Pooling of blood (sequestration) in organs resulting in enlargement:

1. Splenomegaly
2. Hepatomegaly

- Organ damage due to vessel blockage:

1. Heart (cardiomegaly) with weakened heart valves and heart murmur
2. Lungs, kidneys, liver, and spleen malfunction and failure
3. Extremities: avascular necrosis due to vascular blockage resulting in skeletal deformities (hip, shoulder, lordosis, and kyphosis) and possible osteomyelitis
4. Central nervous system (seizures, paresis)
5. Eyes: visual disturbance, possible progressive retinal detachment and blindness

- Growth retardation may also be noted

Diagnostics:

- Low RBCs.
- Sickled cells noted per stained blood smear.
- Sickle-turbidity test (Sickledex).
- Hemoglobin, hematocrit, and platelets.
- Hemoglobin electrophoresis: separation of blood into different hemoglobins to determine the form of hemoglobinopathies (hemoglobin defects).
- Newborn screening for SCA: detects hemoglobin defects early.
- Pulse oximetry and blood gases may reveal hypoxia in severe anemia.
- Acidosis may result in a decreased serum pH level.
- Electrolyte imbalance may be noted due to acidosis.

Treatment:

- Hydration to thin blood and decrease sickling and vascular blockage.
- Minimize infection; antibiotics may be ordered, vaccines recommended to avoid meningitis, pneumonia, and other infections.
- Oxygen supplement to decrease tissue ischemia.
- Pain medication: oral or intravenous analgesics such as opioids.
- Electrolyte replacements may be ordered to correct imbalances.
- Blood replacement with packed cells if anemia is severe.
- Bed rest with mild range of motion during episodes.

NCLEX!!! Avoid cold and cold compresses with increased vasoconstriction and pain.

Nursing Interventions:

- Pain control; fear of addiction is not the issue during a crisis.
- Fluid intake: Monitor intravenous fluids closely to avoid fluid overload.
- Intake and output to regulate volume and monitor kidney function.
- Rest periods during the day to avoid fatigue.
- Mild range of motion to retain mobility

Health Teachings:

- Teach proactive care to prevent episodes/crisis:

1. Adequate fluid intake to prevent dehydration
2. Avoiding infection or early treatment
3. Moderate activity and adequate rest to avoid fatigue and hypoxia

- Early signs of impending crisis: splenic palpation to detect sequestration
- Stress need for immediate care if there are signs of crisis.
- Genetic testing and counseling:

1. Explain that SCAis an autosomal recessive condition requiring the gene from both parents.
2. Encourage testing of siblings to allow for childbearing planning.
3. Explain that each pregnancy when both parents are carriers presents a 25% chance a child will be born with the disease and a 50% chance the child will have the sickle cell trait.
4. Refer for counseling and family planning if additional childbearing is desired.
5. Discuss alternative parenting options (insemination, adoption, etc.).

- Support child and family with emotional responses, grieving, and coping:

1. Allow ventilation of anger, concerns, fears, and questions.
2. Support during depression over chronic illness.
3. Provide honest responses regarding care during episodes.
4. Use positive terms and avoid words like “crisis” when discussing vasoocclusive or other problem episodes with the child and family.
5. Encourage child in control of condition and lifestyle needed to avoid episodes and promote maximum development.

V. HEMOPHILIA

Hemophilia is a group of congenital bleeding disorders due to a deficiency of specific coagulation proteins. This condition occurs most commonly in persons of African descent, possibly as a genetic adaptation in trait carriers as protection from malaria.

Hemophilia results most often from a genetic defect and most commonly a deficiency of factor VIII (hemophilia A) or factor IX (hemophilia B, or Christmas disease). However, a third of hemophilia cases occur from gene mutation. The X-linked form of the condition is passed when an affected male (XhY) mates with a female carrier (XhX) producing a 1 in 4 chance of the offspring having a girl or having a boy with the disease, having a female carrier, or having a child without the disease or trait. The female carrier can also be symptomatic.

Assessment:

- Bleeding of varied degrees depending on severity of deficiency:

1. Spontaneous bleeding

2. Bleeding with trauma
3. Bleeding with major trauma or surgery

- Hemarthrosis(bleeding into the joints) in the knees, elbows, and ankles begins with stiffness, tingling, or ache as early sign of bleeding, progressive damage.
- Warmth, redness, swelling, and severe pain and loss of movement.
- Epistaxis (not most frequent bleed).
- Hematomas may cause pain at the site due to pressure.
- Intracranial bleeding can cause changes in neurostatus and progress to death.

NCLEX!!! Bleeding from the mouth, throat, or neck could result in airway obstruction and warrants immediate attention.

Diagnostics:

- History of bleeding with X-linked inheritance evidenced is diagnostic.
- Clotting factor function testing will reveal an abnormality in ability to form fibrinogen or generate thromboplastin:

1. Whole blood clotting time
2. PT
3. PTT
4. Thromboplastin generation test (TGT)
5. Prothrombin consumption test
6. Fibrinogen level

- Pulse oximetry and blood gases may reveal hypoxia in severe anemia.
- Acidosis may result in a decreased serum pH level.
- Electrolyte imbalance may be noted due to acidosis.

Treatment:

- Factor VIII concentrate to replace the missing clotting factor.
- DDAVP (1-deamino-8-D-arginine vasopressin) for mild hemophilia (type 1 or IIA) to increase production of factor VIII.
- Corticosteroids for chronic hemarthrosis, hematuria, acute hemarthrosis.
- Ibuprofen or other nonsteroidal antiinflammatory drug (NSAID) for pain relief
- Epsilon aminocaproic acid (EACA, Amicar) blocks clot destruction.
- Exercise and physical therapy with active range of motion as client tolerates to strengthen muscles around joints.

NCLEX!!! NSAIDs should be used cautiously because they inhibit platelet function.

NCLEX!!! After acute episode, avoid passive range of motion due to possible joint capsule stretching with bleeding. Client should control active range of motion according to pain tolerance.

Nursing Interventions:

- Maintain protective environment to prevent injury to client.

- Monitor closely for signs of bleeding.
- Treat bleeding episodes promptly.
- Apply pressure to nares if nosebleed is noted.
- Minimize crippling due to contractures and joint damage from bleeding:

1. Promote complete absorption of blood from joints.
2. Mild exercise of limbs during confinement to prevent disuse.
3. Encourage regular exercise regimen at home

VI. IDIOPATHIC THROMBOCYTOPENIC PURPURA

A hemorrhagic condition, is an acquired disorder with an unknown cause that is possibly autoimmune in origin. It occurs between 2 and 10 years of age with recovery within 6 months.

Excessive destruction of platelets results in deficiency (thrombocytopenia) leading to bleeding disorders. Bone marrow may be normal with large young platelets noted. The disorder may be acute or chronic.

Assessment:

- Petechiae, or bruising, due to bleeding in superficial skin surfaces.
- Bleeding from mucous membranes.
- Prolonged bleeding from wounds.
- Fatal hemorrhage is rare.

Diagnostics:

- Platelet count <20.000 mm3

Treatment:

- Supportive treatment
- Prednisone
- Anti-D antibody (if client is >1 year of age or <19 years of age)
- Intravenous immune globulin (IVIG)
- Splenectomy (after 5 years of age) for clients with severe chronic ITPto remove risk of hemorrhage followed by prophylactic penicillin and vaccines to prevent influenza, meningitis, or pneumonia

Nursing Interventions:

1. Supportive care:

- Protective environment with padding to prevent injury.
- Limit activity until platelets 50,000 to 100,000/mm3.

2. Client and family teaching:

- Teach to avoid all contact sports.
- Medical examination with any abdominal or head trauma to rule out internal bleeding.

3. Premedicate with acetaminophen 5 to 10 minutes before anti-D antibody infusion and monitor for reaction of fever, chills, or headache. Treat with Benadryl and Solu-Cortef and observe client closely.

NCLEX!!! Teach client and family to avoid using aspirin or NSAIDs for pain management due to effect on platelets.

VII. BETA-THALASSEMIA

Thalassemia is an inherited disorder involving deficiency in production of globin chains in hemoglobin. The beta form of the disorder is the most common and is found most often in persons of Greek, Italian, and Syrian descent. An alpha form of thalassemia is found in people of Chinese, Thai, African, and Mediterranean descent possibly due to genetic mutation because of intermarriages or spontaneous mutation.

Thalassemia is an autosomal-recessive disorder in which the alpha or beta polypeptide chains in hemoglobin Aare impacted. In beta-thalassemia there is a decreased synthesis of the beta chains with an increased synthesis of alpha chains resulting in defective hemoglobin and damaged red blood cells (hemolysis) and resulting anemia. An overproduction of RBCs (immature cells) may result in compensation for the hemolysis. Folic acid deficiency may result from increased demand on bone marrow.

Assessment:

- Anemia with accompanying:
- Pallor.
- Fatigue.
- Poor feeding.
- Progressive chronic anemia: Hypoxia, headache, irritability, precordial and bone pain, and anorexia may be noted.
- Thalassemia minor occurs with trait carrier condition and is nonsymptomatic.
- Thalassemia intermedia manifests with splenomegaly and moderate to severe anemia.
- Thalassemia major (Cooley anemia) is severe anemia.
- Excessive iron storage in organs without organ damage (hemosiderosis) or with cellular damage (hemochromatosis) may be noted.
- Retarded growth; particularly delayed sexual maturation is commonly noted.
- Bronzed complexion: iron-containing pigment may be noted due to breakdown of RBCs and excess iron.
- If untreated, bone changes such as enlarged head and other facial changes may be noted.

Diagnostics:

- RBC count is low.
- Hemoglobin and hematocrit levels are decreased.
- Hemoglobin electrophoresis analyzes the hemoglobin variants and helps distinguish the type and severity of thalassemia.

Treatment:

- Maintain adequate hemoglobin levels to reduce bony deformities and expansion of the bone marrow.
- Provide blood cells to promote growth and maintain activity tolerance:

1. Transfusions of RBCs as needed to keep Hgb >9.5 g/dL

- Deferoxamine (Desferal), an iron chelating agent, with oral vitamin C may be administered to promote iron excretion (may help growth if given early at 2 to 4 years of age).
- Bone marrow transplantation may be done in some children.
- Splenectomy may be done to decrease destruction of blood cells, if severe splenomegaly is noted.

NCLEX!!! After splenectomy, client is at risk for infection and should receive vaccines to prevent influenza, meningitis, and pneumonia in addition to regular immunizations.

Nursing Interventions:

- Promote adherence to treatment regimen.
- Support child during illness and distressing treatments.
- Promote child and family coping:

1. Anticipate adolescent concerns related to appearance.

- Monitor closely for complications of the condition and treatment:

1. Multiple transfusions and iron buildup
2. Infection postsplenectomy

- Genetic counseling:
- Encourage testing of siblings to allow for childbearing planning.
- Explain that each pregnancy when both parents are carriers presents a 25% chance a child will be born with the disease and a 50% chance the child will have the thalassemia trait.
- Refer for counseling and family planning if additional childbearing is desired.
- Discuss alternative parenting options—insemination, adoption, etc.