Hematologic & Cardiac System Outline – Child

I. Iron deficiency Anemia

A. Incidence & Etiology
   1. Caused by inadequate supply of dietary iron
   2. IDA can be caused by any number of factors that decrease the supply of iron, impair the absorption of iron, increase the body's needs for iron, or affect the synthesis of Hgb.

B. Clinical Manifestations
   1. Weight either under or overweight
   2. Pale mucous membranes
   3. Weak/fatigued easily

C. Diagnostics
   1. Decreased Hgb, Hct, MCV, MCH/ MCHC
   2. Decreased serum iron concentration
   3. Stool for occult blood

D. Therapeutic Management
   1. Dietary counseling, Resources (WIC)
   2. PO supplements, Parenteral iron (IM or IV route)
   3. Transfusions – for severe anemia
   4. Nursing Implications-Prevention

II. Sickle Cell Anemia

A. A group of diseases termed hemoglobinopathies in which normal Hgb is partially or completely replaced by abnormal sickle Hgb (HbS)

B. Incidence- Inheritance=Autosomal recessive disorder, affects many African Americans.

C. Pathophysiology-
   1. Abnormal sickle shaped RBC's- RBCs are rigid and sticky, not flexible
   2. Abnormal shaped cells get stuck in the small vessels

D. Clinical Manifestations-
   1. Varies in severity and frequency
   2. Crises – the most acute symptoms occur during periods of exacerbations
      a. Sequestration Crisis- Spleen enlarged and blood pools in the spleen causing shock state
      b. Aplastic Crisis- bone marrow becomes tired and can't produce any more RBC's
      c. Hyperhemolytic Crisis- RBC destruction is at a greater rate than RBC production
      d. Vasocclusive Crisis- painful episode causing pain and ischemia

E. Other complications
   1. Chest syndrome- pulmonary infiltrates like pneumonia
   2. CVA- Sickled cells block major blood vessels in brain = cerebral infarction

F. Diagnosis
   1. Newborn screening- is mandatory in U.S. as a standard screening
      a. Sickledex screening- finger or heel stick test. Quick results.
      b. Hgb electrophoresis- a fingerprinting looking for normal and abnormal types of hemoglobin in the blood. Done if sickledex screening is positive.

G. Treatment
    1. Goals
        a. Prevent sickling phenomena
        b. Treat crisis
    2. Prevention
        a. Rest, keep hydrated, keep oxygenated, keep healthy
    3. Medical Management of a Crisis
        a. Pain control
        b. Hydration/Electrolyte replacement
        c. Bedrest
        d. Oxygen
    4. Treat infections
    5. Transfusion to treat anemia, exchange transfusions prn
    6. Splenectomy
    7. Vitamins

H. Nursing Considerations
    1. PCN
    2. Maintain adequate hydration
    3. VS monitoring
    4. Support
    5. Genetic counseling

I. Prognosis
    1. Majority of deaths occur from overwhelming infection- a chronic illness

III. Rheumatic Fever

A. Definition- inflammatory disease
    1. Reaction to group A beta hemolytic streptococcus infection
    2. Often follows attack of pharyngitis, tonsillitis, scarlet fever, strep throat, impetigo

B. Incidence- Children and adolescents

C. Etiology- 2-6 weeks after child had infection, got better, then gets sick again

D. Clinical Manifestations
    1. Carditis
        a. Inflammation of the heart. Can have chest pain/systolic murmur or muffled heart sounds
        b. Rheumatic heart disease- affects layers of heart, mostly mitral valve
    2. Polyarthritis
        a. Swollen, red, hot, painful joints that are painful
        b. Favors large joints- knees, elbows, hips, shoulders, wrists
    3. Erythema marginatum
        a. Erythematous macular rash with clear center and wavy well demarcated border
        b. Can be on chest/abdomen/extremities- (non-itchy)
    4. Chorea
        a. Sudden aimless irregular movements of extremities
        b. Speech disturbances & facial grimacing
        c. Muscle weakness
    5. Other Manifestations
        a. Small (0.5 – 1.0 cm) nodules non-tender over bony prominences

b. Arthralgia
c. Fever

E. Diagnostics
1. ESR/CRP elevated plus two major manifestations
2. Positive rapid strep or recent strep infection

F. Therapeutic Management
1. Bedrest especially if carditis is severe
2. PCN – drug of choice
3. Anti-inflammatory medications- Aspirin/Naproxen

G. Prophylaxis against recurrence infections that can cause RHD and further damage
1. Monthly IM PCN injections for 5-10 years or until age 21 depending on case

H. Prognosis/ Sequelae
1. Follow medically for at least 5 years

IV. <u>Kawasaki Disease</u> - (Mucocutaneous Lymph Node Syndrome)

A. Definition: Kawasaki disease causes inflammation in the walls of medium-sized arteries throughout the body, including the coronary arteries, which supply blood to the heart muscle. Kawasaki disease affects lymph nodes, skin, and the mucous membranes inside the mouth, nose, and throat.

B. Etiology- Unknown but affects young children and boys more than girls

C. Clinical Manifestations

1. Difficult to Dx as it mimics other disorders/diseases. 3 Phases of disease:
- <u>Acute Phase</u>-fever, red eyes, red pharynx, strawberry tongue, red dry cracked lips
- <u>Sub-Acute Phase</u>-Fever will resolve, peeling skin large sheets to fingertips/toes
- <u>Convalescent Phase</u>- Resolution of symptoms slowly

2. Cardiac Involvement- complications which can cause arrhythmias, coronary artery aneurysms and scarring and fibrosis of coronary arteries due to ischemia.

C. CDC Diagnostic Criteria

Fever lasting longer than 5 days plus at least four of the following:
1. Bilateral conjunctiva inflammation without exudate
2. Oral mucous membrane changes – strawberry tongue, cracked lips
3. Extremity changes- redness and edema, then peeling of hands/feet
4. Rash- maculopapular
5. Cervical lymphadenopathy

D. Medical Management
1. High dose IV IG- Immunoglobulin
2. ASA salicylate therapy
3. Assess Cardiovascular Status
4. Monitor fever
5. Promote skin integrity
6. Maintain adequate nutrition and hydration

V. <u>Mononucleosis</u>

Definition: Acute, self-limiting viral infectious disease that is common among adolescents. Fever, exudative pharyngitis, lymphadenopathy, hepatosplenomegaly, and increased lymphocytes.

A. Pathophysiology/ Etiology
1. Epstein Barr Virus (EBV)
2. Believed to be transmitted by direct intimate contact with oral secretions
3. Contagious- contact with saliva

4. Period of communicability- indeterminate, virus can be excreted for months

B. Clinical Manifestations
1. S&S vary and mimic many other conditions
2. Early signs- general malaise, chills, low grade fever, loss appetite
3. Full blown disease Triad- fever, sore throat, cervical adenopathy
4. Common features- splenomegaly, palatine petechiae, erythematous trunk rash

C. Diagnostics
1. Mono spot test- quick, inexpensive, easy to perform blood test

D. Therapeutic Management
1. Supportive treatment
2. Analgesics for pain and fever
3. Bed rest
4. Activity Restrictions
5. Corticosteroids used in complicated cases

E. Complications- Ruptured spleen, neuro involvement, secondary infections

F. Prognosis- Self limiting. Usually will get better with supportive treatment.

VI. <u>Acute Lymphocytic Leukemia (ALL)</u>

A. Overview- Most common childhood cancer and accounts for 80% of all childhood leukemia. Malignant disease of bone marrow, blood, and lymphatic system

B. Pathophysiology
1. Healthy children have bone marrow that makes blood stem cells
2. These blood stem cells usually mature and become myeloid and lymphoid cells
3. Myeloid cells mature to become RBC/WBC/Platelets
4. Lymphoid cells mature to become lymphoblasts
5. In Leukemia-these cells don't mature to become the normal cells they should be, they stay immature and can't function effectively. Usually there was some type of genetic or acquired injury that causes this issue.

C. Causes- Unknown, perhaps environmental factors, prenatal exposure to X-rays,

D. Risk factors- Genetics, Environmental, Immunodeficiency issues, Viral infections

E. S/S- Appear within weeks to months of malignant process
-Fatigue (lethargy), paleness, easy bruising, prolonged bleeding, epistaxis, petechiae, frequent infection, joint pain, bone pain, weight loss, anorexia, fever, lymphadenopathy, hepatomegaly, splenomegaly, mediastinal mass, testicular infiltration

F. Diagnosis
1. Symptoms and H&P presentation
2. Lab work & Bone marrow biopsy
3. Lumbar puncture to look for CNS involvement

G. Treatment
1. Goal of treatment is remission of the cancer
2. Chemotherapy, radiation therapy, chemo with stem cell transplant, targeted therapy
3. 3 phases of Treatment:
a. Induction Therapy Phase
b. Consolidation/Intensification Phase
c. Maintenance (continuation) Therapy Phase
4. Remission- relatively good prognosis for remission with treatment
5. Manage any complications/side effects of treatment

H. Stem Cell Transplant- IV infusion of healthy donor stem cells

I. Follow up- Long term complications such as heart damage, poor school performance, secondary malignancy, sterility/sexual development issues. Good prognosis- 90% survival rate

J. Nursing Care & Nursing Interventions

1. Preparation for tests & procedures
2. Pain control
3. Complications of myelosuppression
4. Bleeding & Anemia
5. Drug toxicity
6. Education & Support

VII. Lead Poisoning

A. Incidence- Poisoning from lead goes back to the early 1900's

B. Causes- almost always lead based deteriorating paint. Other Pathways: Food, Air, Soil, Water, ethnic remedies, collectible toys, pottery, occupation-based causes like construction, painting.

C. Children and Lead

1. Young children absorb about 50% of the lead they are exposed to
2. Do not need to eat paint chips to be exposed- Ingestion or Inhalation/Placental transfer

D. Pathophysiology- can affect any part of the body but renal, neuro and heme systems are most seriously affected

1. Heme System- Lead can interfere with the binding of iron onto the heme molecule= Anemia
2. Renal System- Affects the proximal tubules of the kidney in severe cases
3. Neurological System- The developing brain is the most susceptible and most concerning in kids. Can cause behavioral and cognitive problems.

E. Diagnostic Eval

1. BLL (Blood Lead Level Test) Screening should be done before school if not earlier.
2. Acceptable level is less than 5mcg/dL
3. Levels higher than 5 need family education to stop the exposure

F. Therapeutic Management- Chelation Medications used for removing lead from circulating blood, from organs and tissues. Meds bind to the lead and then is excreted through kidneys

1. Chelation agents such as calcium disodium edetate (EDTA) can cause toxic side effects
2. Usually given IV or IM- can be painful
3. Adequate hydration to be maintained during chelation therapy

G. Nursing Considerations

1. Goal: prevent the child's initial or further exposure to lead.
2. Discharge Planning- medication admin and follow up, referrals PRN

H. Prognosis- Most of the effects are reversible, but could have cognitive behavioral issues