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
- **2.** PO supplements, Parenteral iron (IM or IV route)
- **3.** Transfusions – for severe anemia
- **4.** Nursing Implications-Prevention & teaching

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- 1 in 10 black Americans carry trait. Inheritance=Autosomal recessive disorder.

C. Pathophysiology-
- **1.** Sickle shaped RBC's, shaped like a crescent moon
- **2.** RBCs are rigid and sticky, not flexible
- **3.** Abnormal shaped cells get stuck in the small vessels which slow/block blood flow and 02 to parts of the body

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
- **3.** Hand Foot Syndrome- vascular issues, avascular necrosis, leg ulcers

F. Diagnosis

1. Newborn screening- is mandatory in U.S. as a standard
- **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.** Education
- **2.** PCN
- **3.** Maintain adequate hydration
- **4.** VS monitoring
- **5.** Support
- **6.** Genetic counseling

I. Prognosis
- **1.** Most without symptoms participate in normal activities without restriction
- **2.** Majority of deaths occur from overwhelming infection
- **3.** Chronic illness – potentially terminal outcome

III. <u>Rheumatic Fever</u>

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
- **1.** Children 6-15 years; peak 8 years

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

D. Clinical Manifestations
1. <u>Carditis</u>
   - a. Inflammation of the heart. Can have chest pain/systolic murmur
   - b. Rheumatic heart disease- all layers of heart, mostly mitral valve
2. <u>Polyarthritis</u>
   - a. Edema, inflammation, effusion in joints
   - b. Favors large joints
3. <u>Erythema marginatum</u>
   - a. Erythematous macular rash with clear center and wavy well demarcated border

b. Can be on chest/abdomen/extremities

4. Chorea
   a. Sudden aimless irregular movements of extremities
   b. Speech disturbances
   c. Muscle weakness
5. Subcutaneous Nodules
   a. Small (0.5 – 1.0 cm) over bony prominences
6. Minor manifestations
   a. Arthralgia
   b. Fever

E. Diagnostics
- **1.** ESR/CRP elevated
- **2.** Positive rapid strep
- **3.** Strep antibody titer elevated

F. Therapeutic Management
- **1.** Bedrest
- **2.** PCN – drug of choice
- **3.** Oral salicylates

G. Prophylaxis against recurrence
- **1.** Monthly IM PCN injections **or**
- **2.** Daily PCN **or**
- **3.** Daily dose of Sulfadiazine

H. Prognosis/ Sequelae
1. Follow medically for at least 5 years
2. Must always use antibiotic prophylaxis for dental work, infections, invasive procedures

IV. Kawasaki Disease - (Mucocutaneous Lymph Node Syndrome)

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 also affects lymph nodes, skin, and the mucous membranes inside the mouth, nose and throat.

A. Etiology- Unknown

B. Clinical Manifestations

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

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

C. CDC Diagnostic Criteria

Fever lasting longer than 5 days plus at least four of the following:
1. Bilateral conjunctiva infection and inflammation
2. Oral mucous membrane changes
3. Lower extremity changes
4. Rash
5. Cervical lymphadenopathy

D. Medical Management

1. Gamma Globulin IV high dose
2. ASA

E. Nursing Management
1. Assess Cardiovascular Status
2. Monitor fever
3. Promote skin integrity
4. Maintain adequate nutrition
5. Pain management

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, mucous, sometimes even tears
4. Period of communicability- indeterminate, virus can be excreted for months

B. Clinical Manifestations
1. S&S vary and mimic many other conditions
2. Onset of symptoms 10 days to 6 weeks after exposure
3. Early signs-
4. Full blown disease Triad-
5. Common features- splenomegaly, palatine petechiae, erythematous trunk rash, exudate throat

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
-Uncommon but can be serious
Liver involvement, Neuro issues meningitis, encephalitis, Guillain-Barre
Pneumonitis, Myocarditis, Hemolytic anemia, Thrombocytopenia, Ruptured spleen

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

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

A. Overview
1. Most common childhood cancer
2. Accounts for 80% of all childhood leukemia

B. Pathophysiology
1. Childhood acute lymphoblastic leukemia (also called ALL or acute lymphocytic leukemia) is a cancer of the blood and bone marrow.
2. This type of cancer usually gets worse quickly if it is not treated

C. Causes
-Unknown, perhaps environmental factors

D. Risk factors
-Genetics
-Environmental
-Immunodeficiency

-Viral infections

E. Diagnosis

-H&P

-Bone marrow biopsy

-Lumbar puncture to look for CNS involvement

F. 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

G. Treatment

1. Goal of treatment is remission of the cancer
2. Chemotherapy with multi-agents
3. 3 phases of Treatment:
    a. Remission/Induction Therapy Phase
    b. Consolidation/Intensification Phase
    c. Maintenance Therapy Phase
4. Remission- relatively good prognosis for remission with treatment

H. Complications

-DIC

-Relapse of ALL

-Septicemia

-Hemorrhage

I. Prognosis

J. Bone Marrow transplant – Stem Cell Transplant

K. Follow up

-Long term complications such as heart damage, poor school performance, secondary malignancy, sterility/sexual development issues

L. Nursing Care & Nursing Interventions

Preparation for tests

Pain control

Complications of myelosuppression

Bleeding

Anemia

Drug toxicity

VII. Lead Poisoning

A. Incidence

-Poisoning from lead known since early 1900's

B. Causes

-Cause is almost always deteriorating paint

-Other Pathways: Food, Air, Soil, Water, Culture/ ethnicity Involvement

C. Children and Lead

-Young children absorb about 50% of the lead they are exposed to

-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

3. Neurological System
   - The developing brain is the most susceptible and most concerning
   - Lead toxicity leads to behavioral and cognitive problems
   - Lead encephalopathy can lead to seizures – coma – death

4. Other general symptoms- stomachaches, nausea, vomiting, weakness, metallic taste

E. Diagnostic Eval
1. BLL (Blood Lead Level Test)
2. Screening
   - CDC guidelines (supported by AAP)
   - Universal or targeted screening depending on home state
   - DELAWARE: All children should have BLL before starting school

F. Therapeutic management= Patient teaching. AVOID LEAD!

G. Acute Lead Poisoning
1. BLL > 5mcg/dl=cause of concern and needs teaching to prevent further worsening
2. Chelation treatment likely necessary for a BLL of 45 or greater
   - Treatment used for removing lead from circulating blood and some lead from organs and tissues

H. Chelation Medications
1. Succimer
   a. PO route
   b. SE Include: N,V,D- Loss appetite, Rash, Increased LFT's, Neutropenia

2. EDTA (Calcium disodium edetate)
   a. IV or IM (most often IV) route
   b. Many side effects:

I. Nursing Considerations
1. Goal: prevent the child's initial or further exposure to lead.
2. Discharge Planning

J. Prognosis
- Most of the effects are reversible
- Most serious consequence is effects on CNS
- Lead encephalopathy – permanent brain damage
- Academic problems usually present up to "juvenile delinquency"