IRON DEFICIENCY ANEMIA

• Results from an inadequate supply, intake, and absorption of iron
• Most common type of childhood anemia
• Inadequate iron supply leads to smaller RBCs, a reduction in the number of RBCs and the quantity of hemoglobin, and a decrease in the oxygen-carrying capacity of the blood

Iron Deficiency Anemia

• Causes:
  – Large milk intake (milk is a poor source of iron)
  – Inadequate iron supply at birth (prematurity)
  – Impaired absorption (diarrhea, malabsorption)
  – Rapid growth
  – Inadequate diet (low in iron)
  – Menses
Iron Deficiency Anemia Symptoms

- INFANT
  - Overweight
  - Pale
  - Frequent infections

- Adolescents
  - Pale
  - Fatigue
  - Decreased attention span
  - Light-headed

Iron Deficiency Anemia - Diagnostics

- Hemoglobin below 11 gm.
- Hematocrit below 33
- Decreased serum iron binding concentration
- Decreased serum ferritin
- Increased total iron binding capacity

Priority Nursing Diagnosis

- Knowledge deficit
- Altered nutrition, less than body requirements
- Activity intolerance
- Risk for altered growth and development
Interventions

- Detection
  - Screen at 6, 12, 18, 24 months and adolescent females

- Nutritional counseling
  - Iron-rich foods
  - Decrease milk intake

Interventions

- Iron supplements
  - Begin at 4 months
  - Premature infants at 2 months
  - Give between meals or with citrus juice
    - (Increases absorption)
  - Use a straw or brush teeth immediately after administering to prevent teeth staining
  - Stools will become green-black

SICKLE CELL ANEMIA

- A hereditary hemoglobinopathy primarily affecting African-Americans
- An autosomal recessive condition whereby normal hemoglobin is partially or completely replaced by the sickle-shaped, abnormal hemoglobin S (Hgb S)
Etiology

- SCD is an autosomal recessive disease. If one parent has the Hbs trait and the other parent is normal, there is a 50% chance that each offspring will inherit the trait. Children with the trait are asymptomatic.
- If each parent carries the trait there is:
  a) A 25% chance that their offspring will be normal
  b) A 50% chance that the child will carry the trait
  c) A 25% chance that each child will have the disease

Incidence

- Approximately 8% of African-Americans carry the sickle cell trait
- Among African-American infants 1 in 375 is affected with sickle cell disease

Pathophysiology

- When exposed to diminished levels of oxygen, the hemoglobin in the RBC develops a sickle or crescent shape
  - Cells are rigid & obstruct capillary blood flow, leading to congestion and tissue hypoxia
  - This hypoxia causes additional sickling and extensive infarctions
Figure 44-3
Sickle cell anemia

Many of these red blood cells show an elongated crescent shape characteristic of sickle cell.

Sickle Cell Crisis

- Symptoms of crisis do not usually appear until age 4-6 months
  - Sickling of cells prevented secondary to the high levels of fetal hemoglobin
- Those with sickle cell trait (carriers of the disease) rarely experience crises or symptoms
**Types of Sickle Cell Crisis**

- **Vaso-occlusive Crises (Thrombotic)**
  - Most common type of crisis, painful
  - Caused by stasis of blood with clumping of cells in the microcirculation, ischemia and infarction
  - Hypoxia, low oxygen tension or any condition that increases the body’s need for oxygen may trigger a crisis. Sickling may be triggered by fever, dehydration, emotional or physical stress
  - Signs include fever, pain (particularly in the joints, extremities or back), swollen joints, vomiting, anorexia, positive Homans’ sign, tissue engorgement

**Complications of Vaso-occlusive Crisis**

1. Painful episodes
2. Acute chest syndrome
3. CVA

**Types of Sickle Cell Crisis**

- **Acute sequestration**
  - Blood pooling causing decreased hemoglobin level
  - Signs and symptoms include, enlarged liver and spleen, tachycardia, dyspnea, weakness, pallor, shock.
Complications of Acute Sequestration

• Hypovolemic shock

Treatment of Acute Sequestration

• Blood transfusions to restore circulating volume

Types of Sickle Cell Crisis

• **Aplastic Anemia**
  • Cause by viral infection which ceases blood cell production by the bone marrow
  • Usually last 5-10 days
  • Rarely recurs
  • Signs and symptoms include, pallor, lethargy, headache, weakness, tachycardia, fainting, shock
Types of Sickle Cell Crisis

- Acute chest syndrome
  - Signs and symptoms include: chest pain, cough, fever, abdominal pain

Treatment of Acute Chest Syndrome

- Hydration
- Analgesics (morphine or meperidine)
- Bed rest
- Antibiotic administration
- Vaccination
- Blood transfusion (packed RBC are given to treat the anemia and decrease the number of circulating cells)
Diagnostic Tests and Labs

- Multisystem assessment and history
- Newborns may be diagnosed by hemoglobin electrophoresis of cord blood
- CBC (Hgb, Hct, RBC with morphology, WBC count)
- Chest x-ray
- Sickledex (sickle turbidity test) is a screening test used for children over 6 months of age
- Hemoglobin electrophoresis confirms the diagnosis (it differentiates between those children with the trait and those with the disease)

SCA - Priority Nursing Diagnoses

- Altered tissue perfusion
- Risk for injury
- Risk for infection
- Pain
- Knowledge deficit

SCA - Nursing Interventions

- Maintain adequate oxygenation (administer oxygen as needed, monitor pulse oximeter)
- Administer analgesics around the clock
- Assist hydration: IV fluids, oral fluids
- Promote and encourage rest
- Prevent infection: administer antibiotics as ordered
- Provide emotional support
- Encourage child to wear Medi-Alert band
- Offer genetic counseling to parents
SCA – Client/Family Teaching

• Signs & symptoms of impending crisis
• Signs & symptoms of infection
• Importance of adequate hydration
• Preventing hypoxia from physical and emotional stress

HEMOPHILIA

• A group of bleeding disorders characterized by a deficiency in a specific clotting factor
• The disease is a chronic inherited bleeding disorder
• Hemophilia is without cure and is a lifelong condition

Hemophilia A

• Hemophilia A is the result of factor VIII deficiency
• Accounts for 80% of hemophilia cases
• Inherited as an X-linked recessive trait
  – Female inherits the hemophilia trait from her father, then has a 50% chance of transmitting it to her son
  – 1 in 5,000 male births
Risk Factors

- Male with a family history of hemophilia or unexplained bleeding
- Mother with a recessive gene trait

Hemophilia A - Assessment

- Prolonged bleeding from minor trauma
- Careful history concerning episodes of bleeding as well as history of familial bleeding tendencies
- Hematuria or flank pain
- GI, spinal cord bleeds
- Joint bleeding (hemarthrosis) = cardinal sign (can cause permanent crippling)
- Excessive bleeding from circumcision (if an infant) or any break in skin
- Bruising in various stages
- Decreased hemoglobin or hematocrit
- Increased WBC and platelets
- Nose bleeds and/or oral bleeding
- Abnormally long menstrual flow

Hemophilia - Diagnostics

- Prolonged activated PTT - increased
- Prothrombin time (PT) normal
- Bleeding time normal
- Platelet count normal
- CBC
- Decreased factor VIII levels
- Liver biopsy
- Liver function test to rule out liver disease and other causes of bleeding
- Genetic testing to identify carriers
- Amniocentesis
Priority Nursing Diagnoses

• Risk for injury
• Pain
• Impaired physical mobility
• Knowledge deficit

Nursing Interventions

• Control localized bleeding: pressure to insertion site after needle withdrawal, avoid injections, venipuncture, and invasive procedures as much as possible, elevation, local coagulants

• Monitor for bleeding: assess venipuncture site, joints, urine, stools, NG fluids for occult blood

• Transfuse as needed

• No rectal temperatures

• Manage pain

• If joint involved: immobilization, ice packs, elevation, Physical therapy when bleeding controlled

Nursing Interventions

• Replacement of Factor VIII
  – Administered IV when bleeding occurs
  – Parents taught home administration
  – May be administered prophylactically
  – Risks include AIDS, hepatitis, infections
  – Expensive

• Injury prevention

• Avoid aspirin products
Pharmacology

• Highly purified or recombinant Factor viii concentrate
• Desmopressin (DDAVP) which increases plasma factor viii levels, which increases platelet aggregation
• Regular immunization
• Cryoprecipitate or fresh frozen plasma (FFP) transfusion.

IDIOPATHIC THROMBOCYTOPENIC PURPURA (ITP)

• Acquired hemorrhagic disorder characterized by:
  a) excessive destruction of platelets (thrombocytopenia)
  b) purpura (a discoloration caused by petechia beneath the skin)
• Cause unknown
• Autoimmune hematologic condition
• Most common bleeding disorder in children
• Peak age 2-5 years
• Usually develops 1-3 weeks following a viral infection (URI, measles, rubella, chicken pox)
• As platelet destruction exceeds production, total number of platelets decreases

ITP - Assessment

• Easy bruising with petechiae (particularly over bony prominences)
• Multiple ecchymotic areas (particularly over bony prominences)
• Hematuria, bloody or tarry stools
• Bleeding from mucus membranes, and gums
• Nosebleeds
• Hematemesis, hemarthrosis, hypermenorrhea
• Hematomas over the lower extremities that may result in chronic leg ulcers
• Lab data: decreased platelet count, increased anti-platelet antibodies
• Bone marrow to look for platelet precursors and rule out oncologic disorder
ITP – Priority Nursing Diagnoses

• Risk for infection
• Risk for injury
• Knowledge deficit

ITP - Management

• Medications
  – Steroids to reduce inflammation
  – Intravenous immunoglobulins (IVIG) to reduce the autoimmune problem
  – Packed RBC may be given to replace blood loss in symptomatic children
  – Platelet are seldom given

ITP – Nursing Interventions

• Management mainly supportive-course of disease self-limiting in majority of cases
• Frequent assessment for bruising or active bleeding
• Assess neurological status every shift and PRN
• Monitor platelet counts. No rectal temperatures
• Avoid aspirin products
• Provide soft toothbrush
LEUKEMIA

ALL

AML

Leukemia

• Cancer of the blood-forming tissues
• Most common form of childhood malignancy
• More males than females after age 1 year
• Peak onset 2 to 6 years
• Two types recognized in children
  – Acute lymphoid leukemia (ALL)~80% of all childhood leukemias
  – Most common type in children
  – Most common type affecting children <5 years
  – Peak onset is 4 years
  – More common in Caucasians and boys
  – Acute non-lymphoid myelogenous leukemia (AML) or Acute Nonlymphoid Leukemia (ANLL)
  – Affects all ethnic groups equally
  – No peak age at onset

Leukemia-Etiology

• Genetic factors
• Radiation and chemical factors
• Children with immunodeficiency states
• Exposure to viruses before or after birth
Leukemia - Pathophysiology

- Unrestricted proliferation of WBCs occur
- Bone marrow infiltration crowds out stem cells that normally produce red blood cells and platelets; anemia and thrombocytopenia occur; the WBCs that are produced are immature and cannot fight infection
- Spleen, liver and lymph nodes become infiltrated and enlarged

Leukemia - Pathophysiology

- Central nervous system (CNS) is at risk for infiltration
- Clinical manifestations are directly related to areas of involvement, such as bone pain from marrow proliferation

Consequences

- The three main consequences of leukemia in the child include risk for:
  a) Infection due to neutropenia and the lack of WBC defenses
  b) Anemia related to decreased RBC production
  c) Bleeding due to impaired platelet formation
Clinical Manifestations

- Fever
- Pallor
- Overt sign of bleeding
- Lethargy
- Malaise
- Anorexia
- Large joint or bone pain
**Leukemia - Diagnosis**

- History & physical
- Peripheral blood smear reveals anemia, thrombocytopenia, and **neutropenia**
  - Leukemic blasts may be seen on smear
- Bone Marrow aspiration is the definitive test
  - Normal marrow contains less than 5% blasts
  - Leukemic marrow has much higher percentage, often 60-100% blasts
- Spinal tap

**Leukemia – Priority Nursing Diagnoses**

- Risk for infection
- Risk for injury
- Activity intolerance
- Anxiety
- Risk for ineffective family coping
- Pain

**Leukemia - Treatment**

- Aim of treatment is to induce a remission (less than 5% blasts in the bone marrow)
- Chemotherapy
  - Combinations of drugs are used
  - Several stages
- Possible radiation therapy
Chemotherapy

- **Induction**
  - Goal is to induce a remission
  - Lasts 4-6 weeks
- **Intensification or consolidation**
  - Serves to maintain the remission and/or further reduce tumor burden
- **CNS prophylaxis aids intrathecal medication**

**Chemotherapy**

- **Delayed intensification**
  - Uses additional drugs to target leukemia cells that have survived
- **Maintenance**
  - Lasts 2-3 years
  - Preserves the remission
- **Reinduction** is used for relapses and adds drugs not previously used
- **Bone marrow transplants** are considered after a second relapse occurs

**Complications of Chemotherapy**

- **CNS toxicity or damage**
- **Potential damage to the liver, kidneys, GI tract, heart, lungs, hematopoietic, and immune system**
- **Secondary malignancies**
Leukemia – Nursing Management

- Prepare child/family for diagnostic tests and procedures
- Relieve pain
  - Opioids are titrated to the child's needs
    - Administered around the clock
- Provide emotional support to child and family

Nursing Assessment

- Gentle physical assessment is mandatory
- Observe for bruising and new sites of bleeding
- After chemo is started monitor renal function
- Monitor nutritional status
- Monitor CNS function
Nursing Management

• Prevent complications of myelosuppression
  – Infection
    • Private room
    • Reverse/protective isolation
    • Strict handwashing
    • Restriction of visitors
    • Adequate nutrition
    • Masks

Nursing Management

• Hemorrhage
  – Platelet infusions
  – Avoid skin puncture
  – Meticulous mouth care
  – No rectal temperatures
  – Avoid activities that could cause injury

Special Attention to Renal Function when receiving cyclophosphamide

• A side effect is gross hematuria – hydration with IV fluids to attain specific gravity less than 1.010 decreases the severity of hematuria
• Careful monitoring of I&O
• Specific gravity q8 hrs
• Daily weight
Nursing Management

• If treated in the oncology clinic, instruct parents re: safe med administration and signs and symptoms to report
• Ongoing psychosocial assessment and emotional support are essential
• Referral to support groups and social services may be helpful

Manage Problems of Drug Toxicity

• Anemia
  – Blood transfusions
• Nausea & vomiting
  • Antiemetics before chemotherapy
  – Anorexia
  • Small, frequent feedings

Manage Problems of Drug Toxicity

• Manage problems of drug toxicity
  – Mucosal ulceration
    • Bland diet
    • Soft, sponge toothbrush
    • Frequent mouthwashes
    • Local anesthetics
  – Neuropathy
    • Stool softeners
    • Good body alignment, safety measures
Manage Problems of Drug Toxicity

- Alopecia
  - Warn child and parents of possibility before the hair starts to fall out
  - Recommend cotton head coverings
  - Reaffirm that hair will grow back
- Mood changes

Nursing Diagnoses - Chemotherapy

- Risk for infection
- Risk for injury
- Altered nutrition: less than body requirements
- Risk for altered oral mucous membranes
- Altered body image related to hair loss

Fig. 10-4 A & B. T.G. Sundown patient (A) at age 12 (wearing wig because of hair loss during treatment) and (B) 20 years later with wife and daughter.

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THINKING CRITICALLY: QUESTIONS AND SUGGESTED ANSWERS

Thinking Critically: Sickle Cell Anemia

Method is a 12-year-old African American child with sickle cell disease who is admitted to the hospital with severe abdominal pain. While enroute to the ED, he was history of sudden onset of pain in his legs. He woke up the night before with severe pain in his calf muscles, which he described as a burning sensation. The pain was so intense that he could not bear weight on his legs. He was brought to the hospital by his mother.

Method's blood count revealed hemoglobin S levels of 90%. His blood pressure was 130/80 mmHg, and his heart rate was 110 bpm. He was admitted to the ICU for further observation and monitoring. He was started on intravenous fluids and pain medication. His pain was managed with a combination of acetaminophen and non-steroidal anti-inflammatory drugs (NSAIDs).

The pain is likely related to sickle cell crisis, leading to reduced oxygenation of tissues. Method's pain may also be related to dehydration, which can cause vasoconstriction and localized ischemia. A thorough history and physical examination should be performed to rule out other potential causes of pain.

Sickle cell crisis is a medical emergency that requires prompt treatment. Method's pain was managed effectively with intravenous fluids and pain medication. The importance of maintaining hydration cannot be overstated, as dehydration can exacerbate the symptoms of sickle cell disease.

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