Hematologic and Immunologic Dysfunction

Chapter 49
Pages 1601-1642
Debra Mercer BSN, RN, RRT
Carole L. Mackey, MNSc, PNP, RN

Hematologic Function

• Several tests can be performed to assess the hematologic function.
• Normal Lab Values are listed in appendix F (PP. 1894-1903)
• CBC (shift to the left-common term describing abnormal CBC results)
• Iron
• Coagulation Studies
• Immune status
• H & P
• Comments made by parents/care providers

CBC

• WBC (number)
• WBC Differential (types)
• RBC
• Hemoglobin
• Hematocrit
• Platelet
WBC
- WBC (number)
- WBC Differential (types)
- Neutrophil
- Basophil
- Eosinophil
- Monocyte
- Lymphocyte

Neutrophils
- Immature Neutrophils: (band or rod-like shape)
- Increased Neutrophils
- Acute infection

Basophils
- Histamine (vasodilation)
- Heparin (anticoagulant)
- 1% of leukocytes
Eosinophil

- Normal Value: 1-4%
- Produces Histamine
- Destroys Parasites

Monocyte

- Normal Value: 4 - 6%
- Macrophage

Lymphocyte

- Normal Value: 16 - 45%
- T-cells
- B-cells
- Natural killer (NK) cells
- Increase with Viral infections
Platelets

- Cell Fragments
- Function: Blood Clotting
- 150,000-450,000 platelets in each microliter of blood

Platelet Counts

- Bleeding Disorders
- Bone Marrow Disease
- Thrombocytopenia
- Thrombocytosis

RBC

- Red Blood Cell Count
- Hemoglobin
- MCV
Hemoglobin

- **Male:** 13.8-17.2
- **Female:** 12.1 – 15.1

Hematocrit

- **Male:** 40.7 – 50.3%
- **Female:** 36.1 – 44%

Mean Corpuscular Volume (MCV)

- **Normal:** 80 – 96
- **Macrocytic**
- **Microcytic**
RBC Morphology

- Size
- Shape
- Color

Red Cell Disorders

- Anemia:
  - Reduced red blood cells (RBC’s) or the hemoglobin (HgB or Hb) concentration
  - Oxygen-carrying capacity of the blood decreased
  - Most common hematologic disorder of infancy and childhood.
  - Manifestation of an underlying pathologic process.

Anemia classifications

- Etiology or Physiology
  - RBC
  - Hgb
- Morphology
  - RBC Size
  - Shape
  - Color
Effects of Anemia

- Hemodilution
- Decreased Peripheral Resistance
- Cyanosis
- Growth Retardation
- Delayed Sexual Maturation

Therapeutic Management

- Treat underlying cause
- Reverse anemia
- Blood component
- Substance the blood needs for functioning.
- Supportive care:
  - Oxygen
  - Bed rest
  - Replacement of intravascular volume
  - Prevent complications
  - Support family
Nursing Care Management

- Assessment
- Careful history
- Stool exam for occult blood
- Allow child to play with equipment
- See Nurse alert p.1604 for explanations about blood cells that children can understand.
- Prevent Complications

Iron Deficiency Anemia

- **SYMPTOMS**
  - Brittle nails
  - Anemia
  - Pale skin, palms, inner eyelids
  - Constipation
  - Difficulty swallowing
  - Painful tongue

Iron Deficiency Anemia

- Toddler (12-36 months)
- Adolescents
- Premature infants
- Reduced iron stores
- Inadequate supply of dietary iron.
- Most prevalent nutritional disorder in the United States.
Transfer of Maternal Iron to Fetus

- Last trimester of pregnancy.
- Full-term infants have adequate iron stores for 5-6 months.
- Premature infants have only enough iron stores for 2-3 months.

Therapeutic Management

- Iron fortified formula and cereals
- Do NOT give infants under 12 months of age fresh cow’s milk
- Oral iron supplements (3 months)
- Assess Hgb in 1 month
- Further assessment if Hgb fails to rise

Nursing Care Management

- Instruct on administration of iron.
- Stools turn tarry green color.
- Assess stools at follow-up appointments.
- Diet
Sickle Cell Anemia (SCA)

1

2

Sickle Cell Anemia (SCA)

Genetic counseling can aid couples in making informed decisions about pregnancies

Sickle Cell Anemia (SCA)

TEST EARLY FOR SICKLE CELL

USA 37
Sickle Cell Anemia (SCA)

- African-Americans, Hispanics, and other ethnic groups.
- Incidence in African-Americans is about 8% vs West Africa about 40%

Pathophysiology of SCA

- Features are primarily the results of
  - Obstruction caused by the sickle RBC's
  - Increased RBC destruction
- Vaso-occlusion
- Localized hypoxia
- Tissue ischemia and infarction
- Cellular death.

Clinical Manifestations of SCA

- Sickle Cell Crisis
- Vaso-occlusive crisis (painful episode)
- Acute splenic sequestration
- Aplastic crisis
- Hyperhemolytic crisis
- CVA (reoccur in 70% of cases non-treated)
- Acute Chest syndrome
- Infection
Diagnostic Evaluation of SCA

- Newborns in the US (43 states)
- 80% of Hgb is HgF (false negative)
- Screening:
  - Sickledex (sickle-turbidity test) done by fingerstick in about 3 minutes.
  - Infants > 6 months of age
- If test is +, then Hgb electrophoresis is necessary.

Sickle Cell Crisis

- Anything that increases body’s need for oxygen or alters transport of oxygen
- Trauma
- Infection, fever
- Physical and emotional stress
- Dehydration
- Hypoxia
  - high altitude, poorly pressurized airplanes, hypoventilation, vasoconstriction from hypothermia

Sickle Cell Crisis

- Acute exacerbations that vary in severity and frequency
- Types
  - Vaso-occlusive
  - Splenic Sequestration
  - Aplastic Crisis
Vaso-occlusive Crisis

- Pain in areas of obstruction
- Painful swelling of hands and feet
- Stroke
- Chest Syndrome
- Hematuria
- Priapism

Splenic Sequestration

- Weakness
- Irritability
- Unusual sleepiness
- Paleness
- Big spleen
- Fast heart beat
- Pain in the left side of the abdomen

Aplastic Crisis

Signs and Symptoms
- Paleness
- Fatigue
- "Not feeling good"
- Poor appetite
- Fever
- Low blood count (anemia)
- Recent upper respiratory infection
- Passed out (fainting)
Prognosis

- SCA is a chronic illness with a potentially fatal outcome.
- Majority of deaths < 5 years old resulting from overwhelming infection.
- Physical and sexual maturation are delayed in adolescents.

Therapeutic Management

- **Prevention**
  - Aggressive treatment of infection
  - Possible prophylactic antibiotics from 2 months to 5 years
- **Treat medical emergencies (crisis)**

Nursing Care Management of SCA

- Educate
- Recognize early signs and symptoms or crisis
- Treat the child normal
- Adequate hydration
- Prevention of Pain
Psychosocial Needs

- Coping mechanisms
- Support with genetic counseling
- Financial needs
- Caregiver role strain
- Living with chronic illness in the family

Sickle Cell Ideas

- [www.sickle-psychology.com/NewFiles/children.htm](http://www.sickle-psychology.com/NewFiles/children.htm)

Sickle Cell Anemia (SCA)
Thalassemia

- α-Thalassemia
  - Alpha chains affected
  - Occurs in Chinese, Thai, African, and Mediterranean people

- β-Thalassemia
  - Occurs in Greeks, Italians, and Syrians
  - β is most common and has four forms

Pathophysiology

- Normal postnatal HgB has two β-chains and two β-polypeptide chains.
- Partial or complete deficiency in the synthesis of the β-chain of the HgB molecule.
- Overabundance of erythrocytes are formed.
- Excessive iron form hemolysis is stored in various organs known as hemosiderosis.
Diagnostic Evaluation

• Hematologic studies
• Structurally impaired RBC’s (microcytosis and hypochromia)
• Low H & H
• HgB electrophoresis confirms the diagnosis

Forms of β Thalassemia

• Four types
  – Thalassemia minor
  – Thalassemia trait
  – Thalassemia intermediate
  – Thalassemia major “Cooley’s anemia”

β Thalassemia trait

• Small (microcytic)
• Pale (hypochromic)
• Various shape RBC (poikilocytosis)
β Thalassemia intermedia

- Moderate to Severe anemia
- Splenomegaly
- Bone Deformities
- Blood Transfusions to improve quality of life

β Thalassemia major

- Severe anemia
- Chronic Hypoxia
- Bone Changes
- Severe splenomegaly
- Frequent Blood Transfusions

Therapeutic Management

- Transfusions
- Deferoxamine
- Spleenectomy
- Prevention of infection
Nursing Care Management

- Promote Compliance
- Assist with Coping
- Observe for complications associated with treatments
- Test family members and send for genetic counseling
- Prognosis

Prognosis

- Retarded growth
- Delayed or absent secondary sex characteristics
- Expect to live well into adulthood with proper clinical management
- Bone marrow transplant is potential cure

Aplastic Anemia

**Nadya Kolcova**
Born on August 27th, 2001
Died on February 8th, 2005 from aplastic anemia followed by a fungus infection.
Aplastic Anemia (AA)

- Definition: A condition in which the formed elements of the blood are simultaneously depressed as a result of bone marrow failure.
- Manifestations: Pancytopenia, or the triad of profound anemia, leukopenia, and thrombocytopenia.

AA

- 70% are considered idiopathic
- Other 30% acquired has multiple etiological factors
- Classified as:
  - Moderate
  - Severe

Diagnostic Evaluation

- CBC with pancytopenia
- Bone Marrow Aspiration
Therapeutic Management of AA

- Therapy is directed at restoring function to the marrow.

- Involves two approaches:
  - 1. Immunosuppressive therapy
  - 2. Replacement of bone marrow through transplant

Nursing Care Management

- Prepare child and family for diagnostic and therapeutic procedures
- Prevent complications for pancytopenia
- Support in terms of chronic illness and/or possible poor or fatal outcome
- Treat side-effects of chemotherapy

Defects of HEMOSTASIS

- Process that stops bleeding when a vessel is injured.
- Dysfunction in any of the clotting mechanisms will lead to bleeding or abnormal clotting.
- Clotting depends on three factors:
  - 1. Vascular influence
  - 2. Platelet role
  - 3. Clotting factors
Hemophilia

- Group of bleeding disorders in which there is a deficiency in one of the factors necessary for coagulation of the blood.
- Definitive treatment is based on which clotting factor is deficient. Replacement agents are used.

Types of Hemophilia

- Hemophilia A
  - “Classic hemophilia”
  - Deficiency of factor VIII
  - Accounts for 80% of cases of hemophilia
  - Occurrence: 1 in 5000 males
Types of Hemophilia

- Hemophilia B
  - Also known as Christmas disease
  - Caused by deficiency of factor IX
  - Accounts for 15% of cases of hemophilia

Etiology of Hemophilia A

- X-linked recessive trait
- Males are affected
- Females may be carriers
- Degree of bleeding depends on amount of clotting factor and severity of a given injury
- Up to 1/3 of cases have no known family history
  - In these cases disease is caused by a NEW mutation
Most common Hemophilia’s

- Factor VIII Deficiency: accounts for 80-85% of all hemophilia cases.
- Factor IX Deficiency
- Von Willebrand factor (vWF)

Manifestations of Hemophilia

- Bleeding: mild to severe
- Symptoms may not occur until 6 months of age
- Mobility leads to injuries from falls and accidents
- Hemarthrosis: Bleeding into joint spaces of knee, ankle, elbow leading to impaired mobility
- Ecchymosis

Diagnostic Evaluation

- History of bleeding episodes
- Evidence of x-linked inheritance
- Laboratory Findings
- PTT
- Specific assay procedures
- DNA testing
Therapeutic Management

- Replacement of missing clotting factor
- Prevent crippling effects from joint bleeding
- Corticosteroids
- NSAIDS (cautious use)
- Exercise/Physical Therapy
- Primary Prophylaxis

Nursing Care Management

- Assess and recognize symptoms early
- Prevent bleeding
- Recognize and Control Bleeding
  - Factor replacement and RICE
- Prevent Crippling Effects of Bleeding
- Diet
- Teach children early to take responsibility for their condition
- Support Family and Prepare for Home Health

Prognosis

- Historically, most died by 5 years of age
- Now mild to moderate hemophilia patients live near normal lives
- Gene therapy for future
  - Infused carrier organisms act on target cells to promote manufacture of deficient clotting factor
von Willebrand Disease

- A hereditary bleeding disorder, involving deficiency of von Willebrand factor, a plasma protein, and the carrier for factor VIII.
- von Willebrand factor is necessary for platelet adhesion.
- Transmitted as autosomal dominant trait.
- Occurs in males and females.
- Gene for disease is located on chromosome 12.

Manifestations of von Willebrand Disease

- Easy bruising.
- Epistaxis.
- Gingival bleeding.
- Excessive bleeding with lacerations or surgeries.
- Menorrhagia.
Diagnosis
Laboratory Findings
• Decreased von Willebrand factor levels
• von Willebrand antigen levels
• Decreased platelet agglutination
• Prolonged bleeding time
• PTT may be normal or prolonged

Treatment of von Willebrand Disease
• Infusion of von Willebrand’s protein concentrate
• DDAVP before dental extractions or surgery to prevent bleeding episode
• Aminocaproic acid to treat bleeding in mucous membranes (in some cases)

Idiopathic Thrombocytopenic Purpura (ITP)
• Acquired hemorrhagic disorder characterized by:
  – 1. Thrombocytopenia
  – 2. Purpura
  – 3. Normal bone marrow with normal or increased number of immature platelets (megakaryocytes) and eosinophils.
• Two Forms of the Disease
• 1. Acute, self-limiting
• 2. Chronic condition (>6 months duration)
The acute form is usually seen following an URI, or after childhood diseases such as mumps, measles, rubella, chickenpox or after an infection with parvovirus B19.

Diagnostic Evaluation

- Clinical manifestations
- Platelet count
- Abnormal bleeding time
- Rule out other disorders such as leukemia, lupus (SLE), or lymphoma

Therapeutic Management

- Supportive care as it is self-limiting
- Restrict activity
- Symptomatic treatment
- No therapy for asymptomatic patients
- Spleenectomy (>1 year of ITP)
Nursing Care Management

- Supportive
- Teaching
- Encourage quiet activities
- Do NOT use NSAIDS or ASA
- Do Use Acetaminophen

Disseminated Intravascular Coagulation (DIC)

DIC

- DIC is a secondary disorder of coagulation that occurs as a complication of a number of pathologic processes, such as hypoxia, acidosis, shock, and endothelial damage.
- Can result from many severe systemic diseases such as: congenital heart disease, necrotizing enterocolitis, gram negative bacterial sepsis, rickettsial infection and some severe viral infections.
Pathophysiology of DIC

- Coagulation process is abnormally stimulated
- Excessive amounts of thrombin are generated
- Fibrinolytic mechanisms are activated and cause extensive destruction of clotting factors
Diagnostic Evaluation

- Increased tendency to bleed
- Prolonged PT/PTT and TT
- Profound depressed platelet count
- Fragmented RBC’s
- Depleted fibrinogen

Therapeutic Management

- Control the underlying or initiating cause
- Platelets
- Fresh Frozen Plasma
- Infants may require a Transfusion of fresh blood.
- IV admin of Heparin

Nursing Care Management

- Be aware of the possibility of DIC
- Recognize S/S
- Skills to monitor IV infusion of blood/blood products
Epistaxis

- Septum is highly vascular
- Direct trauma
- Foreign bodies
- Nose Picking
- Mucosal inflammation associated with allergic rhinitis

Neoplastic Disorders

- Leading cause of death in children past infancy.
- Approximately 50% all childhood cancers involve the blood or blood-forming organs.

Leukemias

- A group of malignant diseases of the bone marrow and lymphatic system.
- Complex classification is of utmost importance because of therapeutic and prognostic implications.
- 3-4 cases per 100,000 in white children less than 15 years of age.
- More frequent in males than females after age of 1.
- Peak onset is between 2-6 years-of-age.
Classification

- Morphology
- Cytochemical Markers
- Chromosome Studies
- Cell Surface Immunologic Markers

Pathophysiology

- Unrestricted proliferation of immature WBC's.
- In the acute form the leukocyte count is low.
- Deprive normal cells of nutrients
- Liver and Spleen severely affected.

Chromosomal Studies

- Children with trisomy 21: 20 times' greater risk for developing ALL
- Children with more than 50 chromosomes on the leukemic cells have the best prognosis
- Specific translocations of chromosomes on leukemic cells can affect prognosis
Consequences from Leukemia

- Anemia from decreased RBCs
- Infection from neutropenia
- Bleeding tendencies from decreased platelet production
- Spleen, liver, and lymph glands show marked infiltration, enlargement, and fibrosis

Diagnostic Evaluation

- Suspected by history
- Physical exam
- Peripheral Blood Smear
- Definitive diagnosis is made based on flow cytometry of the cells obtained in the bone marrow biopsy or aspiration.
- Lumbar Puncture

Therapeutic Management

- 4 Phases of treatment
  - Induction Therapy
  - CNS Prophylactic Therapy
  - Intensification Therapy
  - Maintenance Therapy
- Reinduction after Relapse
- Hematopoietic Stem Cell Transplantation
- Prognosis
- Late Effects of Treatment
### Hematopoietic Stem Cell Transplantation (HSCT)

- Donors may be relatives or nonrelatives
- Antigen matched or mismatched
- Peripheral stem cells may be used
- Stem cells from umbilical cord blood

### Risks of HSCT

- Significant risk of morbidity and mortality
- Graft vs. host disease (GVHD)
- Overwhelming infection
- Severe organ damage
- Cure after HSCT: up to 60% or 70%

### Prognosis

- If relapse after HSCT: dismal prognosis
- Identified factors for determining prognosis
  - Initial WBC count
  - Age at time of diagnosis
  - Type of cell involved
  - Gender
  - Karyotype analysis
Nursing Care Management

• Assessment
• Nursing Diagnosis
• Plan of Care
• Implementation of Plan of Care
  – Relieve Pain
  – Prevent Complications of Myelosuppression
  – Precautions in Handling Chemotherapeutic Agents
  – Manage Problems of Drug Toxicity
  – Provide Continued Physical Care and Emotional Support
• Evaluation

Nursing Diagnoses

• Risk for injury related to malignant process, treatment
• Risk for deficient fluid volume related to nausea, vomiting
• Risk for imbalanced nutrition
• Impaired skin integrity
• Altered family processes
• Fear related to diagnosis, procedures, treatments

Desiree Rogers tragically lost her battle with Leukemia in May of 2001 at only two and half years old.
Lymphomas
[www.cancer.med.umich.edu/learn/pwtalking.htm](www.cancer.med.umich.edu/learn/pwtalking.htm)
Talking with Children about Cancer

- Hodgkin Disease
  - More prevalent in 15 to 19 years of age
- Non-Hodgkin Lymphoma
  - More prevalent in children <14 years of age

Hodgkin Disease

- Neoplastic disease originating in lymph system
- Often metastasizes to spleen, liver, bone marrow, lungs, and other tissues

Classification of Hodgkin Disease

- Reed-Sternberg Cells
- Classification A: asymptomatic
- Classification B: temp of 38°C or higher for 3 days, night sweats, unexplained weight loss of 10% or more over previous 6 months
Therapeutic Management

• Radiation
• Chemotherapy (alone or with radiation)
• Prognosis
• Nursing considerations

Non-Hodgkin Lymphoma

• Approximately 60% of pediatric lymphomas are NHL
• Clinical appearance
  – Disease usually diffuse rather than nodular
  – Cell type undifferentiated or poorly differentiated
  – Dissemination occurs early, often, and rapidly
  – Mediastinal involvement and invasion of meninges

Immunologic Deficiency Disorders

• HIV
• AIDS
• Severe Combined Immunodeficiency Disease
• Wiskott-Aldrich Syndrome
Wiskott-Aldrich Syndrome (WAS)

- Baden Thomas Klein was born on February 6, 2003.

Technological Management of Hematologic/Immunologic Disorders

- Blood Transfusion Therapy
- Hematopoietic Stem Cell Transplantation
- Apheresis

Blood Transfusions
Blood Transfusion Therapy

- Verify identity of recipient and verification of donor’s blood group
- Monitor VS
- Use appropriate filter
- Use blood within 30 minutes of arrival
- Infuse over 4 hours maximum

Transfusion Reactions

- Hemolytic: the most severe, but rare
- Febrile reactions: fever, chills
- Allergic reaction: urticaria, pruritus, laryngeal edema
- Air emboli: may occur when blood is transfused under pressure
- Hypothermia
- Electrolyte disturbances: hyperkalemia from massive transfusions or patient with renal problems

Nursing Responsibilities

- Identify donor and recipient blood types and groups before transfusing
- Transfuse slowly for first 15 to 20 minutes
- Observe carefully for patient response
- Stop transfusion immediately if signs/symptoms of transfusion reaction; notify practitioner
Delayed Reactions to Blood Transfusion

• Transmission of infection
  – Hepatitis, HIV, malaria, syphilis, other
  – Blood banks test vigorously and discard units of infected blood

• Delayed hemolytic reaction
  – Destruction of RBCs and fever 5 to 10 days after transfusion
  – Observe for posttransfusion anemia

Hematopoietic Stem Cell Transplantation (HSCT)

• Used to establish healthy cells in both malignant and nonmalignant disease

• Ablative therapy: high-dose combination chemo (with or without radiation) to eradicate unhealthy cells and suppress immune system to prevent rejection of transplanted marrow

Hematopoietic Stem Cell Transplantation (HSCT)

• Stem cells harvested from bone marrow, peripheral blood, or umbilical vein of placenta

• Stem cells given to patient by IV transfusion

• Newly transfused stem cells repopulate ablative bone marrow
Family Concerns

- Difficult decision for HTSC transplant
  - Child faces death without transplant
  - Preparing child for transplant places child at great risk
- No “rescue” procedure if complications follow HTSC transplants
- Nursing considerations

Apheresis

- Definition: removal of blood from an individual; separation of the blood into its components
- Nursing considerations

Apheresis (Greek: “to take away”) is a medical technology in which the blood of a donor or patient is passed through an apparatus that separates out one particular constituent and returns the remainder to the circulation. While being related to it, it is distinct from dialysis, which does not separate directly but rather uses osmosis.
Allogeneic Bone Marrow Transplant (BMT)
• Involves matching histocompatible donor with recipient
• Limited by presence of suitable marrow donor

Stem Cell Transplant

Umbilical Cord Blood Stem Cell Transplantation
• Rich source of hematopoietic stem cells for use in children with cancers
• Stem cells found with high frequency in circulation of newborns
• Benefit of umbilical cord blood is blood’s relative immunodeficiency at birth, allowing for partially matched unrelated cord blood transplants to be successful
Autologous BMT

- Uses patient's own marrow collected from disease-free tissue, frozen and sometimes treated to remove malignant cells
- Has been used to treat neuroblastoma, Hodgkin disease, NHL, Wilms' tumor, rhabdomyosarcoma, and Ewing sarcoma

Peripheral Stem Cell Transplants (PSCT)

- A type of autologous transplant
- Different type of collection from patient
- Stimulate production of high number of stem cells, then collect stem cells by an apheresis machine
- Stem cells separated from whole blood
- Remaining blood cells and plasma returned to patient after apheresis
- Stem cells frozen for later transfusion to patient
Good night, sleep tight, our house shall bleed for adult, you hurt this poor one among.
I didn’t know, your brains too tight,
your fragile skin, so easily breaking.
But you just hoped to let me down.
Then you didn’t want to let me go.
Goodnight, sleep tight, our house shall bleed
Your hearing was so intense.
I screamed you throughout the night,
you were never alone, I was just absent.
But with your death, you let me down.
And you didn’t want to let me go.
Goodnight, sleep tight, our house shall bleed
That night, when I turned, I was alone,
I knew what I faced, your fragile skin,
You learned to hate, your fragile skin.
My heart still breaks, and it’s never done,
why I have to let you go.