Learning Objectives
At the completion of this program, the participants will be able to:
1. Review the incidence and genetics of sickle cell disease.
2. Review the basic pathophysiology of sickle cell disease.
3. Identify the major clinical manifestations of sickle cell disease and the associated pathophysiology, clinical manifestations, treatment, and nursing care for each.

Sickle Cell Disease

Overview
- Definition:
  - A group of hereditary hematologic disorders characterized by a predominance of the abnormal Hb S

Incidence
- Incidence - one of the most common genetic disorders in the U.S.
  - 1 in 12 African Americans in the U.S. carries the trait for SCD
  - 1 in 375 African Americans in U.S. has sickle cell disease
- Most commonly affected populations are from:
  - Africa
  - Mediterranean
  - India
  - Middle East

Genetics
- An amino acid substitution in the sixth position of beta-globin
  - Hb S - valine substituted for glutamic acid
  - Hb C - lysine substituted for glutamic acid

Hemoglobin
- Types - dependent on stage in life and any abnormalities of the genes which regulate hemoglobin
- Composed of 4 globin chains
  - Hb F (Fetal hemoglobin) - 2 alpha + 2 gamma chains
  - Hb A (Adult hemoglobin) - 2 alpha + 2 beta chains

Inheritance
- Autosomal recessive (receive a sickle gene from each parent) or involves coinheritance of HbS with gene for Beta-thalassemia or HbC
- Children born to parents who both have the trait for sickle cell disease:
  - 25% chance of being affected
  - 25% chance of not being affected
  - 50% chance of having the trait
Sickle Cell Trait
- sickle cell trait - one normal Hb A gene and one abnormal sickle Hb gene (i.e. HbSA)
  - 35%-45% sickle hemoglobin
  - sickling occurs only under extreme conditions: hypoxia, hypothermia, acidosis

Types of Sickle Cell Disease
- 4 most common:
  - HbSS
    - homozygous form of SCD; also called sickle cell anemia
  - HbSC
    - a heterozygous form of SCD characterized by HbS and HbC
  - HbSβ+ and HbSβ0
    - Sickle thalassemia diseases: characterized by a gene for sickle cell disease and a gene for beta-thalassemia

Newborn Screening
- All states in the U.S.
- Hemoglobin electrophoresis = screening test
- Malaria - genetic mutation/ protective function

Red Blood Cell (RBC) Characteristics
- RBCs sickle upon de-oxygenation:
  - Stiff, non-pliable resulting in RBC trapping in the vasculature
    - Vaso-occlusion
    - Tissue Ischemia
    - Infarction
  - Un-sickle with adequate oxygen and hydration
  - Reduced life-span of 20 days
    - Chronic hemolytic anemia

RBC Sickling
- Predisposing factors for RBC sickling:
  - hypoxia, dehydration, fever, cold, stress
  - often unpredictable without cause

Pathophysiology of SCD by Systems
Spleen
- Repeated engorgement results in infarction of the spleen and replacement of the functional splenic cells with fibrous tissue by about age 5. This process, called functional asplenia, results in an asplenic condition, which puts the child at high risk for infection.
Splenomegaly is common in early childhood (Hb SS) before functional asplenia occurs. Children with heterozygous forms of SCD may experience chronic splenomegaly.

Liver
Anemia and capillary obstruction cause impaired blood flow to the liver which results in liver failure and necrosis.
Hepatomegaly – present by age 1 and persists throughout childhood
Jaundice – commonly occurs due to the rapid turnover of RBCs and the resultant increase in the bilirubin byproduct
Gallstones – occur in some children because of increased bilirubin breakdown and resultant cholestasis (slow bile flow), which often results in cholithiasis (gall stones). Treatment is a cholecystectomy.

Kidney
Children with SCD are experience hyposthenuria—the inability of the kidneys to concentrate urine. This puts them at risk for dehydration and bedwetting (enuresis).
**Because of hyposthenuria, a specific gravity is an inaccurate measurement of hydration. Instead, ongoing measurement of I&O (particularly intake at 1.25 maintenance fluids status) is key to prevention of dehydration and monitoring of hydration status.

Bone
Bony changes including osteoporosis, widening of the medullary spaces, and thinning of the cortices occur because of the congestion of the bone marrow. As a result lordosis and kyphosis may occur. In addition, chronic hypoxia of the bone puts the child at risk for osteomyelitis (infection of the bone) and aseptic necrosis of the femoral head and shoulder.

Central Nervous System (Stroke or CVA)
CNS is susceptible to occlusion, ischemia, and infarction similar to other areas of the body. The clinical manifestation of this occurrence is called Stroke or CVA. It occurs in as many as 15% of children with SCD resulting in motor impairment, paralysis, and even death.
Treatment is aimed as keeping % Hb S below 30% via transfusion therapy and placing children with history of CVA on a chronic transfusion therapy schedule to suppress the bone marrow and keep the % HbS at low levels. There is high risk of recurrence after the first stroke.
Prevention-focused: Nursing Interventions are aimed at assessment of neurologic changes from baseline
Treatment-focused: PRBC administration to promote low % HbS
Heart
Cardiomegaly and a systolic flow murmur occur due to the stress of chronic hemolytic anemia.

Clinical Manifestations
2 main causes:
- Obstruction by RBCs
- Destruction of RBCs
Vaso-Occlusive Crisis
Acute Chest Syndrome
Infection/Sepsis
Splenic Sequestration
Aplastic Crisis

Major Acute Complications of SCD

Vaso-Occlusion Crisis or Episode (VOC or VOE)
- Overview
  Most common and life-threatening acute complication of SCD
  Majority of cases are managed at home
- Pathophysiology
  - Sickled RBC trapping in microvasculature resulting in:
    - Occlusion
    - Ischemia
    - Necrosis
- Clinical Manifestations
  - Pain
    - Bone, abdomen, chest, back, joints, hands and feet (dactylitis), or penis (priapism)
    - Localized or generalized
    - Hours to days
- Treatment
  - Mainly supportive
  - Folic Acid PO Daily
  - Hydroxyurea
- Nursing Interventions
  - Obtain history of past and present pain management
  - Provide pain assessment
  - Administer pharmacologic interventions
  - Use nonpharmacologic interventions
  - Encourage hydration (IV+PO)
  - Prevent complications
  - Provide patient/family education
    Let’s look at each of these...
Nursing Care of the Child with Sickle Cell Disease: 
Acute Complications
Louise D. Jakubik, PhD, RN-BC

Nursing Interventions
Jakubik et al., 2000

Nursing Intervention #1 for VOC: Obtain history of past and present pain management.
- Describe how you’ve managed this pain episode at home.
- What has been most helpful in alleviating the pain?
- Have you had pain like this in the past? If so, what has helped to alleviate the pain?

Nursing Intervention #2 for VOC: Provide pain assessment.
- Frequency of pain assessment
  - Before each pain management intervention
  - 15-30 minutes after each intervention
  - According to medication dosing interval (minimum) once pain is well-controlled
- Self report
  - Pain = whatever the child says it is (subjective)
  - Pain scales
- Physiologic indicators (vital signs)
- Behavioral indicators (activity level, ease of movement of affected area)
- Pain scales
  - FLACC: nonverbal and young children, usually < 3 years old
  - Faces: 3-7 years old
  - Numeric scale (0-10): 8 years and older (must understand rank order and numeric value)
- Use developmentally appropriate pain scale
- Encourage use of scale at home

Nursing Intervention #3 for VOC: Administer pharmacologic interventions.
- Anti-inflammatory -- NSAIDS: (Motrin, Toradol/Ketorolac)
  - side effects = GI upset
- Narcotics: (Codeine, Morphine, Dilaudid)
  - side effects = drowsiness, nausea/vomiting, respiratory depression, itching, rash
- Tolerance, addiction & dependence
  - Know the difference

Nursing Intervention #4 for VOC: Administer nonpharmacologic interventions.
- Heat (heating pads, shower, bath)
- Massage
- Distraction (involve child life)
- Guided Imagery
Nursing Intervention #5 for VOC: Encourage hydration.
- Goal – to correct and prevent dehydration by decreasing the viscosity of the blood
  - IV + PO = 1 to 1.5 X hourly maintenance fluid requirements
  - IV Fluid should NOT be administered at > 1.5 X IV fluid maintenance requirements due to the risk of fluid volume overload and the resultant development of acute chest syndrome (Rausch & Pollard, 1998)

Nursing Intervention #6 for VOC: Prevent complications.
- Incentive spirometry
  - Age appropriate alternatives
- Cough and deep breathing
- Patient mobility

Nursing Intervention #7 for VOC: Provide patient/family education.
- Medication administration
- Medication side effects
- Nonpharmacologic pain management techniques
- When to call the health care provider
- Participating factors for RBC

- Anticipatory Guidance
  - PRBCs and IV Hydration pre-op (1 ¼ X Maintenance Fluids)
  - Iron chelation to prevent iron overload for kids who receive PRBC transfusion therapy

- Prevention
  - Promote hydration
  - Monitor for dehydration
  - Avoid/minimize risk factors
    - Extreme heat/cold
    - Hypoxia

Dactylitis (hand-foot syndrome)
- swelling, redness, and or pain of hands and feet caused by VOC
- age: 6 months - 5 years (most commonly 6 months - 2 years)

Priapism
- persistent painful erection of the penis caused by RBC sickling in the corpora cavernosa
- treatment:
  - IV fluid
  - Analgesia
  - Needle aspiration via Urology MD
Infection

- Overview
  - Decreased or absent splenic function (autosplenectomy) places the child with SCD at risk for infection.
  - Susceptibility to infection begins at about 6 months of age with congestion of the splenic red pulp.
  - #1 cause of death in children with SCD <5 years old.
  - Most common causative organisms are encapsulated organisms.
    - Streptococcus pneumoniae (S.Pneumoniae)
    - Haemophilus Influenza Type B (HIB)

- Clinical Manifestations
  - FEVER is the first sign of bacteremia in the child with SCD
  - Fever 101.5 or > requires immediate medical evaluation and treatment due to the risk of overwhelming sepsis and death.

- Treatment
  - H&P
  - Lab work: Blood cx, CBC w/diff, other as needed
  - IV Antibiotics

- Nursing Interventions
  - Prompt IV Antibiotics
  - Monitoring for s/s of sepsis
  - Patient and family education
    - Temperature taking
    - When to contact health care provider

- Prevention
  - Penicillin prophylaxis
    - Cooperative Study for Sickle Cell Disease Research
  - PCN PO BID
    - Penicillin 125mg PO BID < 3-5 years old (depending on practitioner)
    - Penicillin 250mg PO BID >3-5 years old
  - Erythromycin PO BID for kids allergic to PCN
  - Routine Childhood Immunizations

Acute Chest Syndrome (ACS)

- Overview
  - Defined as any new infiltrate found on chest x-ray in a child with SCD
  - Clinically similar to pneumonia, but has more serious complications associated with it such as marked acute anemia due to RBC sickling
  - Mortality rate is as high as 25%.
  - There is an increased risk of recurrence of ACS after the first episode.
Pathophysiology
- Causes of ACS include:
  - Infection
  - Intrapulmonary sickling of RBCs
  - Pulmonary edema
  - Atelectasis
  - Pulmonary fat embolism
- Regardless if the particular etiology, ACS results in RBC sickling in the lung.

Clinical Manifestations
- Respiratory distress
- Chest pain
- Hypoxemia (decline in pulse ox below baseline)
- Acute and often dramatic anemia below baseline

Treatment
- Antibiotic therapy
- Pulmonary toilet (chest PT, cough, deep breathing, mobility)
- PRBCs administration for severe anemia (Hb<5-6)

Nursing Interventions
- Monitoring of respiratory status
- IV antibiotic administration
- Judicious use of IVF
- Pain management
- Promotion normal respiratory effort

Prevention
- Incentive spirometry every 2 hours while awake
- Promoting patient mobility
- Adequate pain control (prevents splinting of lungs)
- Avoid overhydration

Splenic Sequestration
- Definition: engorgement of the spleen with RBCs; occurs rapidly over several hours
- Splenic sequestration is a unique problem occurring in patients with SCD causing a trapping of RBCs in the spleen.
- Occurs most frequently in children 2 months to 5 years with HbSS and may occur into adolescence in children with heterozygous (HbSC) SCD.
Clinical Manifestations
- Tachycardia
- Anemia
  - Acute, severe anemia (Hb drop by as much as 2-3 grams)
  - Pallor
  - Fatigue/lethargy
  - Abdominal pain
  - Splenomegaly
  - Cardiovascular compromise (worsening cardiac murmur and/or gallop)

Treatment
- Short Term (Acute)
  - IV Fluid to correct hypovolemia (20 cc/kg Normal Saline IV Bolus)
  - PRBC transfusion to correct anemia (transfuse slowly in small aliquots)
  - Frequent palpation of spleen size (Q 15 min until stable, then 1-2 hours)
  - Frequent CBC’s
- Long Term
  - Splenectomy (...reserved for kids > 5 years of age; must weigh risk of infection vs. risk of splenic sequestration)

Nursing Interventions
- Monitoring of cardiovascular status
  - Monitor heart sounds for worsening of baseline heart murmur or development of a gallop
  - Monitor vital signs for s/s of hypovolemia (tachycardia, hypotension)
  - IVF/PRBC administration
  - Monitoring of spleen size
  - Pain management (abdominal pain in some children with splenic sequestration)

Anticipatory Guidance/Patient and Family Education
- Review signs and symptoms of acute anemia
- Review technique for splenic palpation/measurement
- Review when to call health care provider:
  - Increases in splenic size from baseline
  - Unexplained fatigue, pallor

Aplastic Crisis
- Definition
  - Decreased or absent production of RBC’s usually due to viral infection (parvovirus) resulting in profound anemia
Nursing Care of the Child with Sickle Cell Disease: Acute Complications
Louise D. Jakubik, PhD, RN-BC
Nursing of Children Network Conference 2010

Pathophysiology
- Suppression of the bone marrow leads to cessation of PRBC production and resultant acute drop in Hb
  - Because SCD is a compensated hemolytic anemia where the bone marrow normally overproduced RBCs to maintain the child’s baseline low Hb, suppression of the bone marrow results in an acute and dangerous problem.

Clinical Manifestations
- Clinical manifestations are those seen with acute, profound anemia as discussed in splenic sequestration above.

Treatment
- Serial CBC’s (retic and Hb)
- PRBCs for Hb < 5-6

Nursing Interventions
- Strict isolation (protect other patient’s with SCD)
- CV monitoring for s/s of hypotension (increased HR, worsening murmur, falling Hb)
- PRBC administration if indicated

Anticipatory Guidance/Patient and Family Education
- Review sign and symptoms of acute anemia and when to call the health care provider

Sickle Cell Disease: Home Management Issues
- Baseline Hb
- Spleen palpation
- Review home management of pain
- Pain management and school
- Bed wetting (enuresis)

Sickle Cell Disease: Research
- Hydroxyurea
- Outpatient pain and fever management
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Reference List


