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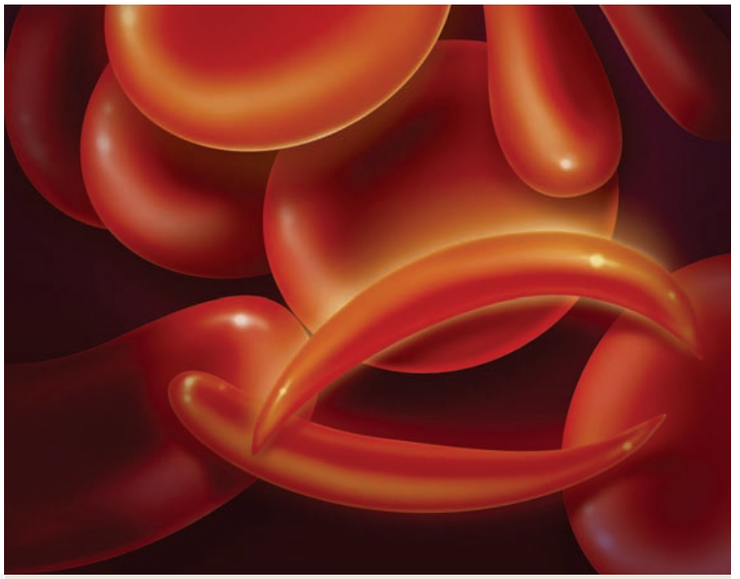
Perinatal Implications of Sickle Cell Disease

Careful planning and execution of nursing care tailored to the childbearing family with SCD can assist in achieving a positive perinatal outcome.

Sickle cell disease (SCD) is an example of a chronic condition requiring complex maternal, fetal, and neonatal nursing care, especially during labor and birth. The purpose of this article is to (1) create an understanding of SCD and its perinatal implications, and (2) describe nursing interventions in caring for the laboring women with SCD.



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Abstract

Sickle cell disease (SCD) affects millions of people across the globe. In the United States, approximately 70,000 to 100,000 people have the disease, and 2 million have the sickle cell trait. SCD occurs once in every 500 African American births, and once in 36,000 Hispanic American births. Women with SCD can have more adverse maternal outcomes such as preeclampsia, eclampsia, preterm labor, placental abruption, intrauterine growth restriction, and low birthweight. Providing comprehensive nursing care to women with SCD is a challenge, particularly during labor and birth, with nursing management aimed at attaining healthy birth outcomes while preventing or treating manifestations of the disease. Labor and delivery nurses are responsible for specific knowledge and care practices for these women, including differentiating the pain of sickle cell crisis from contraction pain and monitoring maternal and fetal oxygenation, as oxygenation is jeopardized in laboring sickle cell patients. Intrapartum nursing care also requires vigilance in the need for emergency cesarean birth. Nursing interventions include symptom management, pain management, ensuring patient safety, and educating patients. Coordination of care and clear communication between the members of the healthcare team, patient, and family are essential elements to ensure a positive outcome for perinatal patients with SCD.

Key Words Chronic disease; Complications of pregnancy; Hematologic diseases; Sickle cell disease.

Understanding SCD

SCD was first described in 1910 by James Herrick, a Chicago, Illinois cardiologist, who discovered oddly shaped red blood cells (RBCs) when he examined a blood sample from a patient who was a West Indian dental student (De, 2008; Friedrich, 2011). The cells were elongated or C shaped, and led Herrick to believe that he had identified a new type of anemia. Several years later, another physician Dr. Victor Emmel termed the peculiar-looking RBCs "sickle cells." In the early 1920s, Dr. Verne Mason originated the term "sickle cell anemia" (Khattab, Rawlings, & Ali, 2006). Today, sickle cell anemia is considered part of a group of inherited blood disorders resulting from a genetic defect in the hemoglobin (Hb) of the RBC. Sickle cell hemoglobin C disease (HbSC) and sickle cell B-thalassemia (HbSBthal) are also part of the group (Zack-Williams, 2007).

Pathophysiology of SCD

Hb is the oxygen carrying protein contained in the RBC. Adult hemoglobin (HbA) is the most common type of Hb and is composed of two α and two β polypeptide chains. Fetal hemoglobin (HbF) contains two α and two γ chains (Kline, 2006). HbF promotes cross-placental oxygen transport in the fetus, as it has a higher affinity for oxygen. It is thus somewhat protective of the potentially hypoxic fetal environment (Rappaport, Velazquez, & Williams, 2004). Postnatally, HbF is gradually replaced by HbA (Frenette & Atweh, 2007).

The Genetics of SCD

SCD is caused by a mutation of the β chain (HbB) of the hemoglobin molecule (HbA) (Porth, 2007). The mutation results in the production of abnormal Hb, hemoglobin S (HbS), associated with three conditions: (1) a most severe form of SCD (HbSS); (2) a milder form, HbSC; and (3) HbSBthal) (Creary, Williamson, & Kulkarni, 2007; Zack-Williams, 2007). The primary genetic defect for HbS occurs on the short arm of chromosome 11 with the substitution of valine for glutamic acid on the sixth amino acid position on the B-globin gene. Persons afflicted with SCD have two copies of the HbS gene. Those with HbSC or HbSBthal have one copy of the C or Thal variant and one copy of HbS mutation (Ashley-Koch, Yang, & Olney, 2000).

SCD is autosomal recessive. For one to have SCD, the person is usually the offspring of parents who are carriers of the gene for the disease (sickle cell trait) (Fisher, 2011). If both parents with sickle cell trait (HbAS) have a child, there is a one in four chance that the child will inherit either the normal hemoglobin gene (HbAA) or sickle cell gene (HbSS). There is a two in four chance of the offspring inheriting sickle cell trait (HbAS). Sickle cell trait is a condition with no anemia or other symptoms.

Morphology

The C shape of the RBCs in SCD is due to the fact that in the deoxygenated state, the solubility of HgS is decreased with the propensity to produce liquid crystals. The crystals cause the cells to form the classic sickle shape (Oteng-Ntim, Chase, Howard, Khazaezadeh, & Anionwu, 2008).

It is this aberrant shape of the RBCs in SCD, which is implicated in the pathology of the disease. The abnormally shaped RBCs are rapidly broken down (hemolyzed) and

destroyed, resulting in their 10 to 20 day life span (normal RBCs have a 90 day life cycle). Consequently, anemia results as RBC production fails to replace the dying cells. The sickle shape also causes RBCs to adhere to the walls of the blood vessels, piling up and causing blockages that damage vital organs and tissues (Ashley-Koch et al., 2000; National Human Genome Research Institute, 2009).

The change in RBC morphology also decreases the oxygen carrying capacity of the cell. This leads to circulatory compromise resulting in the following: vascular stasis leading to impaired circulation; increased blood viscosity and decreased perfusion to organs; and occlusion of the microcirculation leading to tissue hypoxia, infarction, and necrosis (Tanyi, 2003).

Epidemiology

According to the National Heart, Lung, and Blood Institute (2009), SCD of varying types affects millions of people globally. It is common in people who can trace their origins to sub-Saharan Africa, Spanish-speaking regions in the western hemisphere, Saudi Arabia, India, and Mediterranean countries such as Turkey, Greece, and Italy. In the United States, more than 70,000 to 100,000 people have the disease and 2 million have sickle cell trait. SCD occurs once in every 500 African American births, and about once in 36,000 Hispanic American births. One in 12 African Americans has the sickle cell trait (CDC, 2011).

Clinical Manifestations

The clinical manifestations of SCD are caused by two mechanisms: hemolysis and vaso-occlusion. Hemolysis is responsible for the most familiar form of SCD, sickle cell anemia. Vaso-occlusion results in vaso-occlusive or sickle cell crisis, infection, acute chest syndrome, cardiovascular accident, retinopathy, acute splenic crisis, and cholelithiasis (Fisher, 2011; Stuart & Nagel, 2004) (Table 1 for complete description).

Effect of SCD on Pregnancy

The effect of SCD on pregnancy depends upon which manifestation of the disease the woman is experiencing. For example, sickle cell anemia combined with the increased blood volume in pregnancy increases the risks for heart failure should fluid overload occur in therapy for the anemia (Moore, & Martin, 2004). Because of the high cardiac output due to anemia, women with SCD are less tolerant of the increased cardiac output in pregnancy (Creary et al., 2007).

Adverse outcomes in women with SCD include increased medical complications such as infections and thromboembolism, eclampsia, preterm labor, intrauterine growth restriction (IUGR), placental abruption and low birthweight (Sun, Wilburn, Raynor, & Jamieson, 2001; Villers, Jamison, DeCastro, & James, 2008). SCD-related complications also include preeclampsia and urinary tract infections (Sarris et al., 2008). Vaso-occlusive or painful crisis, one of the manifestations of SCD, may be confused with acute abdomen or labor, impeding prompt diagnosis and treatment (Rappaport et al., 2004).

Effect of Pregnancy on SCD

The physiologic changes in pregnancy (hematologic, cardiac, and respiratory) may exacerbate the manifestations of SCD. Rappaport et al. (2004) enumerate the following problems that can result from those changes:

- a profound drop in Hb levels due to the combination of anemia of SCD and the mild anemia caused by pregnancy hemodilution;
- cardiac compromise related to the increased cardiac output of pregnancy;
- acute chest syndrome due to the increased thoracic measurements, elevation of the diaphragm, and the influence of progesterone on the respiratory system, which causes increased airway conduction and total pulmonary resistance;
- exacerbation of vaso-occlusive or sickle cell crisis (Murray & McKinney, 2008), commonly precipitated by acidosis, dehydration, cold temperatures, altitude, stress, fatigue, and infections (Rappaport et al., 2004). Fatigue, dehydration, stress, and infections are commonly experienced in pregnant women, thus the concern for exacerbation of SCD crisis.

Perinatal Management

Management of women with SCD during the perinatal period includes treatment of the underlying disease manifestations. Early and continuous prenatal care is needed to safeguard the fetus/infant during the antepartum, intrapartum, and postpartum periods from potential SCD and pregnancy complications. Toward this end, prenatal visits for the first and second trimester should be scheduled every 2 to 4 weeks (Oteng-Ntim et al., 2008). The postpartum visit is scheduled as usual, unless there are complications.

Intrapartum Care for Women With SCD

Labor and birth requires thoughtful care planning and implementation for the woman with SCD. The nurse not only provides typical labor and birth care but also is vigilant in attending to potential problems resulting from SCD treatment and complications. Anemia, fluid overload, and pulmonary edema are examples of potential problems (Cunningham et al., 2010). The following case study illustrates the critical thinking and clinical skills utilized by nurses caring for these women:

Case Study

History

M.P. was a 20-year-old AA G1 P0 who presented to OB triage at 37 weeks gestation. She had a history of

Table 1. Selected Sickle Cell Disease Manifestations

Etiology/Symptoms	Lab tests	Treatment	Maternal side effects	Fetal side effects
Sickle cell anemia				
Hemolysis and destruction of RBCs RBC 2-3 million Pallor, fatigue, dyspnea	HgB electrophoresis Genetic screening CBC Reticulocyte count Urine culture	Hydroxyurea Folic acid Transfusion (simple or exchange)	Potential transfusion reaction High iron levels Isosensitization	Not given in pregnancy: teratogenic
Vaso-occlusive crisis				
Microvessel occlusion at one or more sites, with inflammation. When severe, known as sickle cell crisis. Acute episodes of pain in any body organ or joint. Can last from hours to days, or chronically for weeks	HgB electrophoresis CBC Reticulocyte count	Opiates Analgesia Anti-inflammatory agents Hydration Transfusion Oxygen Heparin	Pain Sedation Thromboembolism Narcotic dependence	Hypoxia Preterm birth Drug exposure
Acute chest syndrome				
Sickled cells trapped in lung Cough, fever, chest pain, worsening anemia	Chest X-ray Arterial blood gases CBC ECG V/Q scan Blood and sputum cultures Pulse oximetry Pulmonary function testing Brain MRI	Heparin Oxygen Transfusion Antibiotics Respiratory therapy Mechanical ventilation	Bleeding Pneumonia Pulmonary fat embolism	Preterm birth Hypoxia
CVA				
Narrowed, blocked blood vessels Headache, slurred speech, paralysis	MRI Clotting times	Surgery Transfusion therapy	Impaired mobility Impaired speech Impaired vision	Hypoxia Preterm birth
Cholelithiasis				
Increased breakdown of RBCs and increasing blirunbin excretions Pain, fever, nausea, vomiting	Serial LFTs Uric acid CBC, platelets 24-hr urine protein	Cholecystectomy IV hydration Antibiotics Pain management	Risk of anesthesia Infection Surgical complications	Preterm birth Effects of anesthesia
Infection				
Streptococcus Pneumonia, Hemophilus influenza, E-Coli, and others Elevated temperature, chills, perspiration	Specific cultures: blood, urine, sputum	Antibiotics IV hydration Antivirals?	Side effects of specific antibiotic Specific effects on affected organ function	Infection Preterm birth Hypoxia
Retinopathy				
Diminished retinal circulation Salmon spots, retinal degeneration, blindness	Opthalmoscopic and slit lamp microscopic retinal examination	Focal laser surgery	Transient pain	None
Acute Splenic Sequestration Crisis (ASSC)				
Clogging of abnormal RBCs in spleen Anemia, lethargy, weakness, pallor, hypovolemic shock, abdominal fullness/pain, tachycardia	CBC Blood count, ECG	Blood transfusion, removal of the spleen Blood/fluid replacement	Hypovolemia, transfusion reaction	Preterm labor/birth, hypoxia, effects of anesthesia

Note. CBC = complete blood cell count; ECG = echocardiogram; LFT = liver function test; MRI = magnetic resonance imaging; RBC = red blood cell.
Source: Castagnola and Fioredda (2003); Dauphin-McKenzie, Giles, Jacques, and Harrington (2006); NHBLI (2009); Norris Smith-Whitley and McGowan (2003); Pack-Mabien and Haynes (2009); Rosse, Narla, Petz, and Steinberg (2002); Stuart and Nagel (2004); Walter, Hambleton, and Sergeant (2000).

When sickled cells are trapped in the lung, acute chest syndrome results with typical symptoms of cough, fever, worsening anemia, and pleuritic or nonpleuritic chest pain. Pulmonary arterial hypertension can result from the tissue damage that occurs with acute chest syndrome.

SCD diagnosed when she was 7 years old. M.P. was on hydroxyurea prior to her pregnancy, but the medication was discontinued when she became pregnant. Hydroxyurea increases the production of HbF, thereby averting RBC sickling. It is teratogenic in animals and is commonly discontinued in pregnancy (ACOG, 2007). As a young child and teenager she had several transfusions and exchange transfusions. None were required during this pregnancy. M.P. is taking prenatal vitamins, iron, and folic acid.

M.P. was comanaged by her hematologist, nurse midwife, and obstetrician. Her pregnancy labs were within normal limits and she currently did not have anemia. She did have several sickle cell crises during her pregnancy. The crises resulted in hospitalizations for pain management.

Objective Data

M.P. was 5 cm/100%/-2 station upon admission. Her membranes were intact. Her contractions were 5 minutes apart and regular. M.P. complained of contraction pain 7/10, and also was having pain in both legs. An epidural was administered for labor pain management. Maternal O_2 saturations ranged from 85% to 90% on room air. The fetal heart rate baseline was 130, with repetitive late decelerations. Continuous electronic fetal monitoring was instituted. M.P.'s husband and family are with her.

Subjective Data

M.P. stated that she had been nauseated since her contractions began and unable to keep fluids down for the past 6 hours. M.P. complained of contraction pain occurring every 7 minutes and lasting 10 seconds.

Admission Assessment

When women with SCD present in labor, the nurse makes a rapid and accurate assessment of what is happening. The major question to be answered is whether the patient is in sickle cell crisis. This requires careful consultation with the obstetric team for pain assessment and astute evaluation of the continuous fetal monitor strip to differentiate contractions from abdominal pain due to vaso-occlusive events. If the woman is experiencing crisis, the goals of treatment are twofold: (1) manage pain and, (2) ensure adequate fetal oxygenation during the crisis. Differentiation of vaso-occlusive pain from contraction pain is difficult, as labor pain is often located in the abdomen, and abdominal pain is frequent in sickle cell crisis. The fetus frequently has variable or late decelerations related to poor oxygenation.

Nursing Goals

Nursing care goals for M.P. are to: Coordinate the activities of the healthcare team; implement interventions leading to an effective labor and safe birth; monitor mother and fetus for complications of labor, birth, and SCD; provide effective pain management; and initiate labor and educational support.

Nursing Interventions

M.P. was started on oxygen @10 L/min per face mask. She was positioned on her left side and an IV of LR was started. A bolus of 500 mL of fluid was administered rapidly by the nurse for hydration. Her O_2 saturation improved slightly to 95%. M.P.'s HGbS was elevated. The nurse continued to note late decelerations with each contraction. M.P. was experiencing a vaso-occlusive crisis concurrently with labor. This was identified by the nurse through careful assessment. The patient's increased HGbS, leg pain, decreased oxygen saturation, and the late decelerations of the fetal heart rate all suggested a vaso-occlusive episode. A Foley catheter was inserted and the nurse prepared M.P. for cesarean birth. M.P. was taken to the OR and had a viable infant girl weighing 6 lb 10 oz with Apgar scores 7 and 8.

The operative birth was performed because the fetus was not responding to maternal oxygenation and treatment with IV fluids. Sickle cell patients often have microinfarcts in the placenta that compromise uteroplacental

blood flow, resulting in late decelerations. She was not close to vaginal birth and the fetus was not tolerating the contractions.

This case study illustrates the quick and thorough nursing assessment of maternal and fetal status that should occur when patients are admitted in labor with sickle cell crisis.

Clinical Implications for Intrapartum Care

Coordinating the healthcare team activities surrounding the mother with SCD in labor is paramount to deliver effective care. As part of the healthcare team, the nurse provides nursing interventions for the labor patient aimed at pain management, maternal/fetal safety, and patient education. The overall objective is a healthy outcome for the childbearing family.

Pain Management

Pain associated with SCD manifests itself in diverse ways. Pain is subjective, requiring good communication between the nurse and the patient to assess and manage it. Clarity of communication is particularly important during labor because pain associated with labor may be confused with that of vaso-occlusive crisis. The nurse assesses other parameters such as HGbS levels and oxygen saturation to determine whether the pain is partially due to sickle cell crisis. Pain management during labor includes epidural analgesia, an effective method of labor pain management. If not in labor, the patient with SCD would be given opioids to control pain (National Guideline Clearinghouse, 2009). The nurse caring for the patient with SCD in labor with an epidural monitors for potential complications including bleeding, temperature elevation, and maternal hypotension, thereby decreasing fetal oxygenation (Piotrowski, 2007). Distraction and breathing techniques are also part of the pain management plan as is emotional support of the patient and her family.

Maternal/Fetal Safety Goals

Safe care is so important for all patients that National Patient Safety Goals have been advocated and communicated by The Joint Commission (2010). Safety goals applicable to the care of the labor patient with SCD are

- eliminate transfusion errors related to patient misidentification;
- improve the effectiveness of communication among caregivers;
- improve the safety of using medication;
- accurately and completely reconcile medications across the continuum of care;
- and reduce the risk of healthcare associated infections (The Joint Commission, 2010).

The nurse facilitates these goals by cross-checking transfusions (if ordered), and medications, communicating clearly with all members of the healthcare team, verifying and reconciling medications with physician, nurse midwife, pharmacist, patient, and other involved caregivers. Infection and risk of infection are avoided by using scrupulous hand hygiene, monitoring for signs and symptoms of infection, and avoiding exposure to infected individuals.

Patient Education

Labor may be an ideal time for discussion and support of alternative methods of pain management, such as distraction, relaxation, or a combination of both. The immediate postbirth period is appropriate for teaching techniques of breastfeeding, and where to obtain information regarding the compatibility of breastfeeding with pain and other medications used for SCD. Opioids that are typically used for sickle cell pain management can cause sedation in the newborn and require close monitoring of the breastfeeding mother. Folic acid, iron, and hydroxyurea are compatible with breastfeeding (ACOG, 2007). This also presents an opportunity for mention of available breastfeeding classes. The postpartum period provides other teachable moments that include contraceptive choices, breastfeeding support, and emphasis on the importance of keeping appointments for postpartum and SCD follow-up care. Alerting parents to the needs or potential problems of the newborn awaiting diagnosis with SCD is also part of the teaching plan. Both parents need to be taught information about genetic screening and testing, the signs and symptoms of SCD, the treatment of SCD and how to arrange future medical care for their infant.

Several organizations provide information on SCD during the childbearing period (Online). It is an obligation for the nurse to review sites for accuracy and currency and to convey that information to the patient.

Coordination of Care

The nurse communicates with the patient and family to determine needs and to solicit contributions and concerns regarding the medical and nursing management plans. Fostering clear communication between the patient/family, nursing staff, specialty physicians, midwives, advanced practice nurses, and other patient care stakeholders ensures participation by all to deliver comprehensive patient care.

Conclusion

The nurse has a central role to play in helping women achieve a safe birth in the face of challenges posed by a chronic condition such as SCD. Careful planning and execution of nursing care tailored to the childbearing family with SCD can assist in achieving a positive perinatal outcome. ✣

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ONLINE

American Pain Foundation: Patient information for managing pain.
www.painfoundation.org

Sickle Cell Disease Association of America: Information on Sickle Cell Disease and links to educational opportunities, pain management, news, and research.
http://sicklecelldisease.org

March of Dimes: Information on Sickle Cell Disease and Pregnancy.
www.marchofdimes.com/pregnancy/birthdefects_sicklecell.html

Centers for Disease Control: Statistics, causes, symptoms, treatments, illness, prevention. "Sickle Cell Disease: What you should know".
www.cdc.gov/ncbddd/sicklecell

U.S. Department of Health and Human Services, National Institutes of Health. National Heart, Lung, and Blood Institute. Information on Sickle cell anemia, causes, prevention, genetics, newborn testing.
www.nhlbi.nih.gov/health/dci/Sca/SCA_Whatis.html