

Sickle Cell Disease in Pregnancy: Active Nursing Management

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Abstract

Sickle cell disease is considered as a major complication and risk factor for perinatal morbidity /mortality. Literature document that most pregnancies complicated by sickle cell are likely to result in live birth, but the consequences of influence of the disease for the pregnancy/newborn remains a significant concern for health care providers worldwide. According to the bibliography obstetrical-fetal risks are due to the metabolic demands, hypercoagulable state, and vascular stasis associated with pregnancy characterized normally for blood cells to be able to carry oxygen to the growing fetus. With sickle cell anemia, the abnormal red blood cells and anaemic characteristics of the disease physiopathology may result in lower amounts of oxygen going to the developing baby with negative outcome for the future newborn.

Research review studies agreed that access of the pregnant client to a multidisciplinary care team knowledgeable about sickle cell disease and high-risk obstetrics can significantly decrease fetomaternal morbidity and mortality. Example: decreases in spontaneous miscarriage, in perinatal death rates and lowered incidence of preterm labour. Active prenatal management include: education; genetic counselling and prenatal diagnosis for couples at risk; improving nutritional status; vaccination for disease prevention, and early detection of bacterial infection.

***Objective** of this study was to explore active nursing management of the pregnant women with sickle cell disease, including education, treatment and nursing intervention.*

Method: use of English Literature review current through: Jun 2017, Data were searched using MEDLINE, EMBASE, PUBMED and COCHRANE Systematic Reviews.

Keywords: *Sickle Cell Disease, complication, fetomaternal risk, active nursing management.*

Introduction

Sickle cell disease (also called sickle cell anemia) is an inherited blood disorder that affects red blood cells. People at risk for inheriting the gene for sickle cell, are descended from people who are or were originally from Africa or parts of India and the Mediterranean. Population mobility has spread the sickle cell gene through Europe, Asia, Americas and the Caribbean. It means that millions of people have Sickle Cell Disease worldwide. They are either carriers of or have the sickle cell trait. Carriers are usually asymptomatic and have a low percentage of sickle hemoglobin (HbS). Two parents who are carriers can both pass on the sickle cell trait to their offspring, resulting in SCD. There is a 50% chance with each pregnancy for the child of two sickle cell carriers to be born with the sickle cell trait, and there is a 25% chance for the child to be born with SCD.

The disease has been declared by WHO (2006) as a major world health problem. The sickle cell gene mutation causes the body to produce abnormal haemoglobin. In sickle cell disease, the haemoglobin clumps together, causing red blood cells to become stiff and develop a C-shaped ("sickle") form. These red blood cells can block blood vessels, reducing blood flow, which limit adequate oxygenation to many parts of the body. This contributes to the severe pain experienced as a sickle cell crisis and both short-term and long-term organ damage. The disease also makes patients more susceptible to infections as the spleen may be damage decreasing the individual ability to fight infection.

For some pregnant women, a less percentage of studies show no change in their disease during pregnancy, while others may have a worsening of the disease, resulting from or in many cases as the pregnancy accelerates sickle cell complications surface.

Risks to the woman

Sickle cell disease is associated with:

- Increased incidence of perinatal mortality
- Premature labour
- More acute painful crises during pregnancy
- Increase in spontaneous miscarriage
- Recurrent antenatal hospitalisation
- Maternal mortality
- Delivery by caesarean section
- Infections (especially urinary tract infection) during pregnancy
- Thromboembolic events
- Ante partum haemorrhage
- Increased risk of pre-eclampsia
- Pregnancy-induced hypertension
- Postpartum infection
- Increased numbers of cases with acute chest syndrome
- Increases the risk of blood transfusion reactions
- Increased incidences of thromboembolism and pulmonary infarct, which are usually fatal
- Heart enlargement and heart failure from anemia
- Vision problems.

Risks to the baby

Sickle cell disease is associated with fetal complications such as:

- Premature birth
- Fetal growth restriction or IUGR (intrauterine growth retardation)
- Increase the likelihood of fetal distress
- Chance of their baby being affected by Sickle cell disease
- Birth defects
- Low weight babies
- Stillbirths.
- Newborn death
- Newborn with Severe anemia.
- Neonatal jaundice.

Nursing interventions

Goals of active nursing intervention related to management of sickle cell disease in pregnancy.

- Improve obstetric and neonatal outcomes.
- Give emotional support and measures to alleviate symptoms associated with disease manifestations.
- Enhance patient knowledge for better understanding and cooperation.
- Minimize complications
- Comprise a multidisciplinary approach.
- Promote and maintain a safe environment during pregnancy, labor, delivery, and the postpartum period.
- Enhanced patient sense of self-esteem and power.

- Active Nursing interventions for pregnant patient with sickle cell anemia should start at the client first antenatal visit.
- Early and regular prenatal care allows healthcare provider to keep a close monitoring on the disease condition and on the health of developing baby.
- Antenatal care should be provided by a multidisciplinary team including an obstetrician and midwife with experience of high-risk antenatal care and a haematologist with an interest in Sickle cell disease condition.
- A head to toe assessment should be performed at each visit to discard signs of worsening anemia, joints swelling, appropriate fundal high according to gestational age.
- Assess for fetal movement and normal fetal heart rate.
- Assess for any sign and symptoms of infection.
- Antenatal education is a significant resource in increasing knowledge for the pregnant client, her partner and family involve, about the disease process, with beneficial results for a healthy pregnancy and baby.

The nurse must teach the patient about situations that can precipitate a sickle cell crisis and steps to help prevent or diminish such crises example:

- Keep warm.
- Maintain adequate hydration.
- Avoid stressful situations.
- Educate patient on the importance of maintain a healthy diet.
- Encourage patient to take folic acid, vitamins and iron supplements.
- Enhance good patient-nurse communication and encourage patient to report any changes.
- Educate patient to maintain proper hygiene to avoid infections.

Studies have also demonstrated an increase in the incidence of urinary tract infection and asymptomatic bacteraemia so is recommended a urinalysis to be performed at each antenatal visit and midstream urine should be sent for culture and sensitivity monthly. Blood pressure and weight should be checked at each visit to monitor for signs of preeclampsia.

Ultrasound scanning during pregnancy

Serial growth scans allow early detection of fetal growth restriction and hence aid appropriate timing of delivery to reduce perinatal mortality and morbidity

- Women should be offered a viability scan at 7–9 weeks of gestation.
- Women should be offered the routine first-trimester scan (11–14 weeks of gestation) and a detailed
- Anomaly scans at 20 weeks of gestation.
- serial fetal biometry
- Growth and amniotic fluid monitoring scans every 4 weeks from 24 weeks of gestation.
- *32 weeks and advance, growth and well-being scans.*

This includes

- **Nonstress tests:** measure fetal heart rate
- **Biophysical profile tests:** monitor fetal movements, muscle tone, and breathing movements, etc.
- **Doppler sonography:** monitor blood flow from the placenta to the fetus.

Nursing intervention should focus on

- Managing Pain
- Preventing and Managing Infection
- Promoting Coping Skills
- Monitoring and Managing Potential Complications
- Promoting Home and Community Based Care
- Medications

This includes

- Daily folic acid and prophylactic antibiotics (if not contraindicated).
- Drugs that are unsafe in pregnancy should be stopped.
- Iron supplementation should be given only if there is laboratory evidence of iron deficiency.
- Low-dose aspirin 75 mg once daily from 12 weeks of gestation in an effort to reduce the risk of developing pre-eclampsia.
- Prophylactic low-molecular-weight heparin during antenatal hospital admissions. For the associate risk with risk of venous thromboembolism.

Managing pain

Sickle cell disease (SCD) is associated with chronic haemolysis and painful episodes, during pregnancy, SCD may become more severe, and pain episodes may happen more often; usually happen in the bones, joints, abdominal organs. They can last a few hours, few days, or last for weeks. Sickle cell painful crisis is the number one cause of recurrent hospitalization. It must be treated with medications that are safe to use during pregnancy.

Women admitted with sickle cell crisis should be looked after by the multidisciplinary team, involving obstetricians, midwives, haematologists and anaesthetists. A detail assessment should be rapidly carried out to rule out medical complications requiring intervention such as ACS, sepsis or dehydration.

- Dehydration and electrolyte imbalance caused by vomiting, diarrhoea or pyrexia should be corrected with the administration of intravenous fluid.
- Initial investigations should include full blood count, reticulocyte count and renal function. Other investigations will depend on the clinical scenario but may include blood cultures, chest X-ray, urine culture and liver function tests.
- Oxygen therapy may be required if baseline less than 95% for supplementary therapy and to prevent fatal hypoxia. Sickle cell disease can lead to severe placental damage which decreases the transfer of blood rich in oxygen and nutrient to the fetus.

The World Health Organization analgesic ladder should be used, starting with paracetamol for mild pain; NSAIDs can be used for mild to moderate pain between 12 and 28 weeks of gestation. Weak opioids such as co-dydramol, co-codamol or dihydrocodeine can be used for moderate pain, and stronger opiates such as morphine can be used for severe pain. Morphine or diamorphine can be given by the oral, subcutaneous, intramuscular or intravenous route depending on the woman's preference and local expertise. Parenteral opiates can be given by intermittent bolus or patient-controlled administration systems.

Pethidine should be avoided because of the risk of toxicity and pethidine-associated seizures in patients with SCD. While women are receiving parenteral opiates, they should be nursed in an area where they can undergo hourly observations. Assessments of pain score, sedation score and oxygen saturation should be performed. Opiate morphine constricts the blood vessels in the placenta and so may be harmful to the fetus. Nurses must monitor continuous fetal activity to rule out fetal distress. And provision of social, physical and psychological support to alleviate symptoms associated with chronic pain.

According to the literature review sickle cell pain crisis on third trimester are more likely to last for a longer time or until after delivery. Which required a longer stay in hospital, in most cases until the baby is born.

- The nurse must monitor patient for signs of worsening condition:
- A fever higher than 101° F
- Difficulty breathing
- Chest pain
- Abdominal oedema
- A severe headache

- A sudden feeling of weakness
- Seizures
- Sudden vision loss

Blood transfusion during pregnancy

A systematic cohort review indicated that there is insufficient evidence to draw conclusions about the role of prophylactic transfusion in pregnancy to decrease the incidence of maternal painful crises. Routine prophylactic transfusion is not recommended during pregnancy for women with SCD. If acute exchange transfusion is required for the treatment of a sickle complication, it may be appropriate to continue the transfusion regimen for the remainder of the pregnancy. Risks associated with transfusion, include alloimmunisation (the formation of antibodies to red cell antigens), delayed transfusion reactions, transmission of infection and iron overload.

Blood transfusion therapy should be given only

1. To compensate for anemia with symptoms of impending cardiac failure
2. To provide a prophylactic “top-up” transfusion before a caesarean section
3. Emergency transfusion for acute anemia (< 5 g/dL hemoglobin)
4. Twin pregnancy
5. Previous history of perinatal mortality
6. Septicaemia,
7. Acute renal failure
8. Acute chest syndrome
9. A recent neurologic event, hypoxemia,

Randomized studies have shown that many patients, when closely monitored, had well-tolerated anemia, regardless of the level of hemoglobin, and that many were able to complete their pregnancy successfully without transfusion, regardless of the route of delivery.

Recommendations

1. Preconception care

- Amplify the role of family planning and health promotion in the communities to provide education to all women in childbearing age about Sickle cell disease effect- outcome in pregnancy. And the importance of have the haemoglobinopathy status of their partner before becoming pregnant.
- Promote counselling about reproductive options, planning and contraceptive choice.
- Advice the couple to assist genetic counselling.
- Folic acid (5 mg) should be given once daily both preconceptual and throughout pregnancy.

2. Antenatal care

- Identify the client as a high risk and refer couple to high risk pregnancy clinic.
- Remit patient to a high – risk clinic to be evaluate by a multidisciplinary team including an obstetrician and midwife with experience of high-risk antenatal care and a haematologist with an interest in SC.
- Promote screening programme to ensure that screening tests are offered by 8–10 weeks of pregnancy by Primary care or maternity services.
- To monitor and prevent worsening of the condition.
- Iron supplementation should be given only if there is laboratory evidence of iron deficiency.
- Referring to type of delivery is important to maintain proper temperature on delivery room to avoid hypothermia, acidosis and hypoxemia. And to prevent vaso-occlusion and joint pain
- Care could be improved by more specialist centres and specialist nurses or midwives.

- Guidelines need to be created for all healthcare professionals to improve management of these women during pregnancy.
- Refer to genetic counselling and promote partner screening testing to determine presence of HbS.

Conclusion

The detail of appropriate prenatal care and perinatal management for these patients is still a matter for debate in developed countries. However nowadays outcomes have improved significant for pregnant women with Sickle Cell Disease and newborns thanks to medical/obstetric advancement and early-vigilant nursing care, with a good chance of having a safe and healthy pregnancy. Different studies agreed for the need of a dedicated multidisciplinary health care for pregnant women and newborn with SCD for crucial improvement of the disease condition and prevention of short/long term complications. Optimal management during pregnancy should be directed to education/counselling about healthy diet, prenatal vitamins, folic acid supplements and B vitamins, prevention of dehydration, also avoidance of precipitants such as a cold environment and excessive exercise, that can trigger pain crises, and to prevent chronic organ damage, and optimization of fetal health with the goal of minimizing early maternal mortality.

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Anexo#1

Nursing Diagnosis	Expected Outcome	Nursing interventions	Rationale	Evaluation
<p>1. acute pain related to Intravascular sickling with localized stasis, occlusion, and infarction/necrosis due to deprivation of oxygen and nutrients and accumulation of noxious metabolites</p> <p>Evidenced by Localized, migratory or generalized pain, described as throbbing, gnawing, or severe affecting peripheral extremities, bones, joints, back, abdomen, rated 8 to 9 in pain scale. And headaches recurrent/transient and Facial grimacing,</p>	<p>1. Patient will verbalize relief or relief/control of pain within 6h after interventions</p> <p>2. Patient will Demonstrate relaxed body posture, have freedom of movement, be able to sleep/rest appropriately</p> <p>Within 6hr after interventions.</p>	<p>1. Monitor strict vital sign every 4hr such as blood pressure, temperature, heart rate, oxygen saturation and assess pain for location, duration and intensity using pain scale of 0-10.</p> <p>2. Observe nonverbal pain cues such as: gait disturbances, positioning of the body, guarding behaviour, facial grimacing, and reluctance to move. Using patient's subjective description of pain and pain rating on a pain scale to guide the use of analgesic agents.</p> <p>3. Administer analgesics per mouth/intramuscular every 6-8 hours as indicated by doctor according to World Health Organization analgesic ladder.</p> <p>4. Initiate intravenous fluid administration N/S 0.9% one litre every 8 to 12 hours to maintain adequate body temperature and correct or prevent dehydration.</p> <p>5. Apply warm compress to</p>	<p>1. Sickling cells potentiates cellular hypoxia causing infarction of tissues resulting in pain these usually localize in the back, ribs and limbs, may last for days. Alteration in v/s relate to worsening of the condition and severity of pain. Physiological manifestation of acute pain increase BP and RR and cause tachycardia.</p> <p>2. Nonverbal cues may aid in evaluation of pain and effectiveness of therapy since pain is unique in each patient.</p> <p>3. Analgesics reduce pain and promote rest and comfort.</p> <p>4. Dehydration increases sickling/ vaso-occlusion and</p>	<p>In 6hr after nursing intervention patient reported /shows Relief of pain. Decreased incidence of crisis. Enhanced sense of self-esteem and power. Absence of complications.</p>

<p>narrowed/self-focus</p>		<p>affected joints and other painful areas. 6. Provide support and carefully position or massage affected extremities until swelling diminishes. 7. Encourage ROM exercises. 8. Educate patient about pain management and alternative pain relieve measures such as relaxation or distraction techniques and breathing techniques. 9. encourage patient and significant others active participation in patients care.</p>	<p>corresponding pain. Fever triggers painful sickle cell. 5. Warmth causes vasodilatation and increases circulation to hypoxic areas. 6. To reduce edema, discomfort and risk of injury, especially if osteomyelitis is present. Massage help reduce muscle tension and promote blood circulation. 7. Prevents joint stiffness and possible contracture formation. 8. Cognitive behavioural pain management may reduce reliance on pharmacological means of pain control. This also enhances patient sense of control. 9. Involving patient and significant others in care help in establishing individualized treatment.</p>	
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Anexo#2

Nursing Diagnosis	Expected Outcome	Nursing interventions	Rationale	Evaluation
<p>2. Ineffective (specify) Tissue Perfusion related to Vaso-occlusive nature of sickling, inflammatory response Evidenced by Changes in vital signs: diminished peripheral pulses/capillary refill, general pallor Hb >5 g/dl. Restlessness, Tingling in extremities, intermittent headache and bone pain.</p>	<p>Patient will Demonstrate improved tissue perfusion as evidenced by stabilized vital signs, strong/palpable peripheral pulses, adequate urine output, absence of pain; normal capillary refill; skin warm/dry; nail beds and lips of natural pale, pink color</p>	<p>1. Monitor vital signs: Assess pulse points for rate, rhythm, and volume. Assess for hypotension, rapid, weak, and thready pulses, and increased or shallow respirations. 2. Assess skin for pallor, cyanosis, coolness, diaphoresis, and delayed capillary refill. 3. Monitor changes in level of consciousness, reports of headache, dizziness, development of sensory and motor deficits (hemiparesis or paralysis), and seizure activity. 4. Maintain adequate fluid intake and monitor urine output. 5. Assess the lower extremities for skin texture, ulcerations,</p>	<p>1. Accumulation and sickling in peripheral vessels may lead to complete or partial blockage of a vessel with diminished perfusion to surrounding tissues. This can lead to shock. 2. Changes reflect diminished circulation and/or hypoxia potentiating capillary occlusion. 3. Changes may reflect diminished perfusion to the central nervous system (CNS) due to ischemia or infarction. 4. Dehydration causes increase in sickling and occlusion of capillaries other than hypovolemia or decrease in blood volume. Decrease renal perfusion may indicate vascular occlusion. 5. Sickling of blood can cause reduced peripheral circulation and often leads to dermal changes and Occlusion of blood</p>	<p>After 8 hours of nursing intervention patient show sign of improvement evidenced by vital sign within normal range. No sign of worsening skin pallor or cyanosis. Normal level of consciousness, no headache, and no dizziness reported and no seizure activity noted for the shift. Urine output of 450 mls over 8 hr shift. No sign of worsening or pitting edema</p>

Anexo#3

Nursing Diagnosis	Expected Outcome	Nursing interventions	Rationale	Evaluation
<p>3. Deficient Knowledge related to Information misinterpretation Unfamiliarity with resources Evidenced by Verbal/nonverbal cues of anxiety Questions, request for information, statement of misconceptions</p>	<p>. Patient will Verbalize understanding of disease process, including symptoms of crisis, potential complications. Patient will Verbalize understanding of therapeutic needs.</p>	<p>1. Educate patient about signs and symptoms of the disease for which medical attention should be sought, e.g.: Urine that appears blood tinged ; Indigestion, persistent vomiting, diarrhoea, excessive thirst; Severe joint or bone pain; Severe chest pain, with or without cough; Abdominal pain; gastric distress following meals; Fever, swelling, redness, increasing fatigue/pallor, dizziness, drowsiness. 2. Assist patient to strengthen coping abilities, e.g., deal appropriately with anxiety, get adequate information, and use relaxation techniques. 3. Teach patient about situations that can precipitate a sickle cell crisis and steps to take to prevent or diminish such crises (e.g., keep warm, maintain adequate hydration, and avoid stressful situations). 4. Review patient's current diet, reinforcing the</p>	<p>1. To increase patient knowledge about disease facilitating prompt recognition of worsening condition and access to medical care. 2. Increase knowledge decrease anxiety level and enhance cooperation with medical/nursing interventions. 3. Provides knowledge base from which patient can make informed choices. 4. nutrition is essential because of increased demands placed on bone marrow e.g. folate and vitamin B12 Folic acid supplements are frequently ordered to prevent aplastic crisis 5. Frequent monitoring of CBC is required because of narrow margin between efficacy and toxicity (neutropenia, anemia, and thrombocytopenia).oral hygiene limits opportunity for bacterial invasion or sepsis. Detects development</p>	<p>Patients demonstrate good understanding about disease process and complications as evidenced by decrease level of anxiety. And full cooperation during procedures.</p>

		<p>importance of diet including liver, green leafy vegetables, citrus fruits, and wheat germ. Provide necessary instruction regarding supplementary vitamins such as folic acid.</p> <p>5. Encourage patient to have routine follow-ups, e.g.: Periodic laboratory studies, e.g., CBC; Biannual dental examination; Annual ophthalmologic examination.</p> <p>6. Discuss genetic implications of the condition. Encourage partner to seek testing to determine presence of HbS.</p> <p>7. Explore concerns regarding childbearing and family planning and refer to community resources and obstetrician knowledgeable about sickle cell disease, as indicated.</p>	<p>of sickle retinopathy with either proliferative or non-proliferative ocular changes.</p> <p>6. Hereditary nature of the disease with the possibility of transmitting the mutation may have a bearing on the decision to have children.</p> <p>7. Provides opportunity to correct misconceptions/present information necessary to make informed decisions. Pregnancy can precipitate a vaso-occlusive crisis because the placenta's tortuous blood supply and low oxygen tension potentiate sickling, which in turn can lead to fetal hypoxia.</p>	
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