



Sickle Cell Anemia: An Updated Review for Nursing Interventions

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Abstract

Background: Sickle cell anemia (SCA) is a genetic blood disorder caused by a mutation in the hemoglobin gene, leading to abnormal hemoglobin (HbS) that distorts red blood cells into a sickle shape. This impairs cell flexibility and leads to vaso-occlusion, causing a range of acute and chronic complications. SCA predominantly affects populations in Africa, the Middle East, and India. Management of this condition requires a multidisciplinary approach, with nurses playing a crucial role in symptom management, pain control, and prevention of complications.

Aim:

This review aims to provide updated insights into nursing interventions for managing sickle cell anemia, emphasizing the role of nursing professionals in enhancing patient outcomes through comprehensive care strategies.

Methods: This review synthesizes recent research on the pathophysiology, clinical manifestations, risk factors, and medical management of sickle cell anemia. It explores the nursing diagnosis, including common symptoms such as severe pain, fatigue, and infection risk, and highlights interventions for managing vaso-occlusive crises, pain, hydration, and psychosocial stressors. The management approaches discussed integrate both pharmacological and non-pharmacological strategies.

Results: The findings indicate that nurses play a pivotal role in monitoring vital signs, managing pain, providing hydration, and administering antibiotics to prevent infections. Key nursing interventions include assessing pain levels, ensuring hydration, and monitoring complications like acute chest syndrome, stroke, and infections. Nurses also engage in promoting patient education and supporting psychosocial well-being, which are crucial in improving long-term outcomes.

Conclusion: Effective nursing interventions are integral to the holistic care of patients with sickle cell anemia. A collaborative approach, focusing on symptom management and preventive care, significantly improves patient quality of life. Comprehensive nursing care can prevent complications, reduce hospitalizations, and enhance overall patient outcomes in individuals with sickle cell anemia.

Keywords: Sickle Cell Anemia, Nursing Interventions, Vaso-Occlusive Crisis, Pain Management, Hydration, Infection Prevention, Patient Education.

1. Introduction

Sickle cell anemia is the most severe variant of sickle cell disease, representing the homozygous condition for hemoglobin S. This genetic disorder predominantly affects populations in Africa, the Middle East, and parts of India, particularly in regions historically endemic to malaria. Under normal physiological conditions, hemoglobin remains in a soluble state. However, in individuals with sickle cell disease, hemoglobin undergoes precipitation into insoluble crystalline forms. This phenomenon leads to the deformation of red blood cells (RBCs) into a sickle shape, impairing their functionality and making them susceptible to phagocytosis. These deformed RBCs exhibit reduced lifespan and elasticity, contributing to a cascade of clinical complications [1][2][3].

Nursing Diagnosis

Patients with sickle cell anemia present with a range of clinical manifestations that necessitate vigilant

nursing care. Common symptoms include severe pain localized to the bones, chest, back, and joints, alongside systemic features such as fever, malaise, and anxiety. Cardiopulmonary signs such as tachycardia and tachypnea, as well as shortness of breath, are frequently observed. Complications like gallstones, vision disturbances, leg ulcers, and stroke further underline the systemic impact of this disease. Nurses play a pivotal role in managing these symptoms, prioritizing pain control, monitoring vital signs, and addressing psychosocial stressors. These interventions require a multidisciplinary approach to improve patient outcomes effectively.

Etiology

The genetic basis of sickle cell disease lies in a point mutation within the beta-globin chain of hemoglobin. Specifically, a substitution of adenine (A) with thymine (T) occurs at the sixth codon, replacing glutamic acid with valine in the beta-globin chain. This mutation can affect either one or both chains. When both chains are impacted,

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it results in sickle cell anemia, whereas a single affected chain leads to the milder sickle cell trait. This molecular alteration underpins the pathophysiology of the disease, driving the production of abnormal hemoglobin S, which polymerizes under hypoxic conditions, triggering cell deformation and clinical symptoms [4].

Risk Factors

Sickle cell disease disproportionately affects individuals of African origin, although it also occurs in populations of Middle Eastern, Indian, and Mediterranean descent. Epidemiological studies reveal that approximately 1 in 13 African-American children carries the sickle cell trait, while the disease itself affects 1 in 365 individuals of African descent. In the United States, around 100,000 individuals live with this condition. These statistics underscore the genetic predisposition and public health burden of sickle cell disease, highlighting the need for targeted screening, genetic counseling, and culturally tailored healthcare interventions [5].

Clinical Assessment

Sickle cell disease typically manifests after six months of age, coinciding with a decline in fetal hemoglobin levels that otherwise maintain hemoglobin S in a soluble state. The hallmark clinical feature is the vaso-occlusive crisis, presenting as excruciating pain in various body regions, including the abdomen, chest, joints, and long bones. Some patients experience recurrent crises, while others remain asymptomatic for extended periods. Anemia-related symptoms, such as fatigue, pallor, tachycardia, and palpitations, are also prevalent. Recurrent vaso-occlusion can lead to splenic infarctions, resulting in functional asplenia and increased susceptibility to infections from encapsulated organisms like *Streptococcus pneumoniae* and *Staphylococcus aureus*. These infections often culminate in life-threatening pneumonia and septicemia. Digit vaso-occlusion, or dactylitis, manifests as painful swelling of the fingers, whereas abdominal crises mimic acute abdomen conditions. Acute chest syndrome, characterized by respiratory distress, chest pain, and leukocytosis, is another potentially fatal presentation. Additional complications include splenic sequestration crises, retinal hemorrhages, stroke, gallstones, renal isosthenuria, and priapism. Long-term sequelae include avascular necrosis of the femoral head, requiring surgical intervention, and aplastic crises triggered by parvovirus B19, which exacerbate anemia by suppressing erythropoiesis. Collectively, these complications necessitate comprehensive and individualized care strategies to address the multifaceted challenges of sickle cell disease.

Pathophysiology of Sickle Cell Anemia

Sickle cell anemia results from a genetic mutation in the beta-globin gene, leading to the substitution of valine for glutamic acid at the sixth position of the beta-globin chain. This seemingly minor alteration profoundly impacts hemoglobin's properties. In the deoxygenated state, hemoglobin S (HbS) molecules polymerize, forming long, rigid strands within the red blood cells (RBCs). This polymerization distorts the cells into a characteristic crescent or "sickle" shape, reducing their flexibility and deformability. Normal RBCs are highly pliable, allowing them to traverse narrow capillaries efficiently. However, sickled cells are rigid and prone to becoming trapped in the microvasculature, leading to vaso-occlusion. This blockage impedes blood flow, resulting in ischemia and acute pain

crises, a hallmark of the disease. Moreover, the rigid sickled cells are mechanically fragile and have a significantly reduced lifespan of 10–20 days compared to the typical 120-day lifespan of healthy RBCs. This accelerated hemolysis contributes to chronic anemia and increased bilirubin production, often causing jaundice and gallstone formation.

Repeated episodes of sickling and vaso-occlusion lead to cumulative organ damage. For example, the spleen becomes progressively infarcted due to repeated vaso-occlusion, culminating in functional asplenia. This predisposes patients to infections by encapsulated bacteria such as *Streptococcus pneumoniae*. In the kidneys, sickling in the renal medulla results in isosthenuria, impairing the ability to concentrate urine. Similarly, in the bone marrow, recurrent ischemic injury can trigger avascular necrosis, particularly in the femoral head, leading to severe joint dysfunction. On a systemic level, the disease triggers chronic inflammation. The damaged endothelium promotes the adhesion of sickled cells and leukocytes, exacerbating vaso-occlusion and contributing to a pro-inflammatory state. This chronic inflammation, coupled with endothelial dysfunction, increases the risk of stroke and other vascular complications. In summary, the pathophysiology of sickle cell anemia is driven by the polymerization of HbS under hypoxic conditions, resulting in structural and functional derangements of RBCs. These alterations set off a cascade of events that include vaso-occlusion, hemolysis, ischemia, and chronic inflammation, collectively contributing to the disease's complex clinical manifestations and complications.

Evaluation

In newborns with a familial history of sickle cell disease, early screening is critical. In the United States, screening for sickle cell hemoglobin is mandated for all newborns. Prenatal diagnosis for genetically at-risk fetuses can be performed using advanced techniques such as chorionic villus sampling or amniocentesis [6]. Initial diagnostic evaluation includes a complete blood count (CBC) and peripheral blood smear. Hallmark findings include reduced red blood cell (RBC) and reticulocyte counts, variables mean corpuscular volume, elevated leukocyte levels, decreased erythrocyte sedimentation rate (ESR), and the presence of sickle-shaped cells. The detection of Howell-Jolly bodies serves as evidence of functional asplenia. Hemoglobin electrophoresis is employed to confirm the diagnosis by quantifying hemoglobin types. In sickle cell disease, sickle hemoglobin (HbS) comprises over 90% of total hemoglobin, with fetal hemoglobin making up the remainder. Conversely, a concentration of HbS around 45% indicates the sickle cell trait rather than the disease. Urine analysis is essential to exclude urinary tract infections and to detect isosthenuria, a common renal complication. For patients presenting with acute vaso-occlusive episodes in the absence of a confirmed diagnosis, a rapid sickling test may be utilized, though it cannot distinguish between heterozygous and homozygous forms of HbS. Arterial blood gas analysis is crucial for assessing pulmonary function, especially during acute chest syndrome, with serial measurements aiding in monitoring disease progression. Imaging studies, including chest X-rays and peripheral radiographs, can reveal marrow infarctions or osteomyelitis. Early radiographs may not detect acute bone changes, but subsequent images may show bone destruction and sequestra. Magnetic resonance

imaging (MRI) is invaluable for diagnosing osteomyelitis and avascular necrosis, particularly in the femoral and humeral heads. Technetium-99 scans provide additional diagnostic clarity for osteonecrosis.

Medical Management

The treatment of sickle cell disease focuses on seven primary objectives: managing vaso-occlusive crises, addressing chronic anemia and pain, preventing infections and complications, minimizing stroke risk, and detecting pulmonary hypertension. Effective management relies on both pharmacological and non-pharmacological interventions. Hydroxyurea, an antimetabolite, remains a cornerstone of therapy due to its ability to elevate fetal hemoglobin levels. This prevents HbS polymerization, reducing the occurrence of vaso-occlusive crises and other complications [7][8][9]. Supportive pharmacotherapy includes opioids, nonsteroidal anti-inflammatory drugs (NSAIDs), corticosteroids, antiemetics, tricyclic antidepressants, and antibiotics to alleviate symptoms and manage infections. Vaccination, particularly against encapsulated bacteria, is critical, given the functional asplenia in these patients. Folic acid supplementation is also recommended to prevent macrocytic anemia. Advanced interventions include blood transfusions, especially during aplastic crises or severe anemia, and bone marrow transplantation, which remains the only curative option. Preventive strategies emphasize early detection and comprehensive care to mitigate the disease's systemic impact. In summary, the management of sickle cell disease involves a multifaceted approach combining pharmacological therapies, preventive measures, and advanced medical interventions. This comprehensive strategy aims to enhance patient outcomes and improve quality of life.

Nursing Management

In the management of sickle cell disease, the nursing team plays a pivotal role in providing holistic care for the patient. The initial step is to systematically monitor vital signs, ensuring that any abnormalities are identified early. This is followed by a comprehensive assessment of the patient's pain level, which is essential in managing the painful vaso-occlusive crises commonly seen in this disease. Understanding the degree of pain allows for appropriate analgesic intervention and ensures patient comfort. Nurses should also evaluate the patient's hydration status, as dehydration can exacerbate sickling and contribute to further complications.

An assessment of mental status is crucial in identifying any early signs of complications such as stroke or infection. Oxygenation status must also be carefully monitored, as sickle cell disease can impair respiratory function, particularly in the context of acute chest syndrome. Pain management should include timely administration of pain medications, with a focus on opioids and NSAIDs to alleviate discomfort. Intravenous fluids are essential to maintain hydration and support circulation, reducing the risk of vaso-occlusive episodes. Additionally, the administration of prescribed antibiotics is necessary to prevent or treat infections, especially in patients who may have functional asplenia. Blood transfusions may be required in cases of severe anemia or aplastic crises, helping to restore hemoglobin levels and alleviate symptoms. Bed rest is important for patients experiencing acute pain, as it reduces energy expenditure and helps minimize the risk of complications. Wound care is also essential for preventing infection, particularly in patients

with ulcers or other skin breakdowns. Nurses should ensure that clothing is not restrictive, allowing for proper circulation. Encouraging a well-balanced diet with folate supplementation is key in promoting overall health and managing anemia.

When To Seek Help

Nurses and healthcare providers should be vigilant for signs that necessitate immediate intervention. If the patient experiences increasing shortness of breath, it may signal acute chest syndrome or another respiratory complication requiring urgent care. Unstable or rapidly changing vital signs, such as sudden drops in blood pressure or increases in heart rate, may indicate a life-threatening crisis that demands prompt medical attention. An escalation in pain intensity or fever is also an indication that the patient may be entering a critical phase of illness, which may require adjustment in treatment strategies or additional medications.

Outcome Identification

The ultimate goal of nursing management is to achieve specific, measurable health outcomes that reflect the patient's well-being. Successful management would result in the complete resolution of pain, enabling the patient to maintain daily activities such as eating, drinking, and ambulating without hindrance. Restoration of normal mental status, free from confusion or neurological impairment, is a key indicator of improved health and recovery. Furthermore, the maintenance of normal vital signs is essential in ensuring the patient's stability and preventing further complications. These outcomes are indicators of the effectiveness of nursing interventions and the overall management of sickle cell disease.

Monitoring

Continuous monitoring is crucial to ensuring the patient's condition remains stable and to identifying any emerging complications. Vital signs should be checked regularly, especially in cases of acute vaso-occlusive crises or acute chest syndrome, to track any changes in the patient's condition. Oxygenation levels must be assessed to detect hypoxia or respiratory failure early, while pain intensity should be regularly evaluated to guide appropriate medication administration. Nurses should also monitor hydration status to prevent dehydration, which can exacerbate symptoms of sickle cell disease. Lastly, nutrition plays a significant role in managing the disease, and monitoring dietary intake ensures the patient receives adequate nutrition, including folate supplementation, which is essential in supporting red blood cell production. By continuously evaluating these factors, nurses can optimize care and ensure better outcomes for patients with sickle cell disease.

Coordination of Care

Sickle cell anemia is a severe hereditary disorder characterized by high morbidity and mortality. It typically manifests early in life and often presents various types of occlusive crises, which significantly impair the patient's quality of life. Early screening for sickle cell anemia is mandated at birth in the United States, facilitating prompt diagnosis and treatment. Given that the disease affects nearly every organ system, a comprehensive and interprofessional care approach is essential to ensure optimal management and prevent further complications. As the patient population ages, chronic complications, such as pulmonary hypertension, are emerging with considerable morbidity and mortality. Therefore, the expert consensus emphasizes the need for a multidisciplinary team to manage

sickle cell anemia. This team should include physical therapists, psychiatrists, social workers, nurses, pharmacists, substance abuse counselors, pain specialists, and rehabilitation experts to address the wide array of needs presented by these patients [10][11]. For any patient presenting with fever, an infectious disease consult should be promptly initiated to prevent complications.

Orthopedic involvement is required for patients experiencing hip pain or gait difficulties, while a radiologist is crucial in obtaining bone samples if osteomyelitis is suspected. Ophthalmologic assessments should be regularly performed to monitor potential eye complications, and urologic consultations are necessary to manage priapism. Pharmacists play a pivotal role in ensuring medication adherence, as sickle cell patients often require multiple drugs. Ensuring that patients understand their medications and avoid adverse effects is essential. Nurses should also educate patients on the importance of hydration, vaccination, and consistent follow-up with relevant healthcare providers to promote comprehensive care and enhance quality of life.

Outcomes

The prognosis for most individuals with sickle cell disease is complex and varies significantly. While the goal of treatment is to extend lifespan with minimal morbidity and mortality, many individuals still face premature death despite advancements in care. The morbidity associated with sickle cell disease remains exceptionally high, as nearly all patients experience at least one vaso-occlusive crisis during their lifetime. These patients often face considerable challenges in terms of work capacity due to disability and endure chronic pain, which significantly impairs their quality of life. The primary causes of death in sickle cell patients include acute chest syndrome, pulmonary embolism, and infection, all of which contribute to the high mortality rate. Outside of North America, life expectancy for individuals with sickle cell disease is typically in the 30s or 40s. To address these challenges, several clinical guidelines have been established, such as penicillin prophylaxis for children, routine blood transfusions, and pneumococcal vaccination. Additionally, the use of hydroxyurea has enabled many patients to live longer and healthier lives, offering a significant improvement in overall outcomes compared to the past [12].

Health Teaching and Health Promotion

Effective health education and promotion are essential components of managing sickle cell disease. Nurses should educate patients on the importance of maintaining a healthy diet, including folate supplementation, to support optimal health. Patients should also be advised to avoid triggers that could exacerbate sickling episodes, such as extreme temperatures, dehydration, and physical stress. Vaccination is crucial, particularly against pneumococcus and other encapsulated organisms, to prevent life-threatening infections. Patients should be strongly encouraged to avoid smoking, alcohol consumption, and the use of illicit substances such as cocaine, as these can trigger sickling episodes and worsen the disease. In addition to lifestyle modifications, it is important to teach patients to apply warm compresses to painful areas, as heat can alleviate discomfort during a vaso-occlusive crisis. Hydration is essential to prevent sickling and reduce the severity of symptoms, so patients

should be educated on the importance of maintaining proper fluid intake.

Risk Management

Effective risk management is integral to the care of patients with sickle cell disease. Healthcare providers should remain vigilant for any signs of complications and act swiftly when necessary. Abnormal vital signs, such as elevated heart rate or decreased blood pressure, should prompt immediate evaluation. Respiratory distress or shortness of breath requires urgent attention, as it may indicate the onset of acute chest syndrome or other pulmonary complications. Fever is another red flag, as it could signal an infection, a common cause of morbidity in sickle cell patients, and requires a timely infectious disease consult. Intense pain, particularly if it is persistent or escalating, may necessitate pain management interventions, including analgesia or hospitalization. Low oxygen levels and unresponsiveness should be immediately addressed, as they may indicate severe complications such as stroke or acute chest syndrome. If hypotension occurs, it should be managed with appropriate interventions to prevent shock and ensure the patient's stability. Healthcare providers must act quickly to manage these risks, ensuring the best possible outcomes for patients living with sickle cell disease.

Discharge Planning

Effective discharge planning is integral to improving patient outcomes, particularly in the management of chronic diseases such as sickle cell anemia. One of the most crucial aspects of ensuring favorable outcomes post-discharge is patient education. Patients must be informed about the importance of early intervention, as seeking medical help at the onset of symptoms significantly enhances prognosis. It is essential for patients to promptly seek healthcare assistance if they experience symptoms such as fever, abdominal or chest pain, or persistent headaches, as these may indicate complications that require immediate attention. In addition to seeking timely medical intervention, patients should be educated on lifestyle modifications that can mitigate disease progression and reduce the risk of crises. Specifically, patients should avoid the use of tobacco, alcohol, and illicit drugs such as cocaine, as these substances can exacerbate the disease and trigger vaso-occlusive episodes. Additionally, patients should refrain from seeking care from multiple healthcare providers or institutions without proper coordination, as this can lead to fragmented care and potential risks of miscommunication or inconsistent treatment regimens. To optimize health and prevent exacerbation of symptoms, patients should be encouraged to maintain proper hydration, adhere to a balanced and nutritious diet, and receive recommended vaccinations to prevent infections. Regular follow-up appointments with clinicians are critical to monitoring the disease and adjust treatment as necessary. Furthermore, adherence to prescribed medications, such as hydroxyurea, should be emphasized, as it has been shown to reduce the frequency of vaso-occlusive crises and improve overall health outcomes.

Evidence-Based Issues

Educating patients about hydroxyurea is a critical aspect of evidence-based practice in the management of sickle cell disease. Research indicates that hydroxyurea is effective in reducing the frequency of vaso-occlusive crises, a hallmark complication of sickle cell anemia. As such, patients should be thoroughly informed about the

benefits of this medication and its role in improving both short-term symptom control and long-term health outcomes. Hydroxyurea works by promoting the production of fetal hemoglobin, which helps reduce the sickling of red blood cells, thereby lessening the occurrence of painful episodes and other complications. Given the well-documented benefits of hydroxyurea in managing sickle cell disease, it is essential that patients understand the importance of adhering to their prescribed regimen. Healthcare providers should emphasize that consistent use of hydroxyurea can lead to significant improvements in the patient's overall health and may reduce the need for frequent hospitalizations. Additionally, patients should be counseled about the potential side effects of the drug, as well as the importance of regular monitoring to ensure that the medication is effective and well-tolerated. By incorporating these evidence-based insights into patient education, healthcare providers can empower patients to take an active role in their disease management, ultimately leading to improved outcomes and a better quality of life. [10-15].

Conclusion:

Sickle cell anemia (SCA) is a life-altering genetic disorder with significant clinical implications. Its hallmark feature—painful vaso-occlusive crises—along with other complications such as stroke, acute chest syndrome, and infections, presents a complex management challenge for healthcare providers. The role of nurses in managing SCA is essential to improving patient outcomes, as they are often the primary caregivers responsible for monitoring symptoms, administering treatments, and providing holistic care. Nursing interventions must be comprehensive, addressing not only the physical symptoms but also the psychological and emotional aspects of the disease. Pain management is a priority, as vaso-occlusive crises can cause excruciating pain, requiring careful administration of opioids, NSAIDs, and other analgesics. Hydration is another critical component of nursing care, as dehydration can exacerbate sickling and contribute to further complications. Regular monitoring of vital signs, including respiratory and cardiovascular parameters, is necessary to detect signs of acute complications, such as stroke or acute chest syndrome, at an early stage. In addition to physical management, nurses must address the psychosocial challenges associated with SCA. Patients often experience significant emotional stress due to the chronic nature of the disease, the frequent hospitalizations, and the potential for life-threatening complications. Nurses can provide invaluable support by offering counseling, promoting patient education, and connecting patients with community resources. Education on disease management, preventive care, and the importance of adherence to prescribed treatments is essential for empowering patients and their families. Prevention of infections is also a vital nursing intervention, particularly in patients who have functional asplenia. Vaccinations, regular screenings, and timely administration of antibiotics are necessary to reduce the risk of life-threatening infections. Additionally, blood transfusions and hydroxyurea therapy can help manage chronic anemia and reduce the frequency of crises. In conclusion, the nursing management of sickle cell anemia requires a multifaceted, patient-centered approach. By focusing on pain control, hydration, infection prevention, mental health support, and patient education, nurses can significantly improve the quality of life for individuals living with this debilitating condition. Their involvement in

both acute and long-term care is critical to reducing complications and promoting better health outcomes.

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