CHAPTER **29**

SICKLE CELL DISEASE: Perioperative Management in Podiatric Surgery

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INTRODUCTION

Approximately 1% of the African American population in the US carries the diagnosis of sickle cell disease (SCD).[1] SCD is a general term for patients with some degree of (ß-hemoglobin chain mutation. Over the last decade, the medical community has increased their understanding of the pathophysiology of this disease and therefore, sickle cell disease patients are living longer due to improved medical treatment options. Despite advancements, these patients continue to be a medical management challenge, especially when planning for surgery. It has been well-documented within the medical literature that sickle cell disease predisposes the patient to an increased risk of perioperative complications when compared to the general population. This is primarily based on the multiple potential consequences of vaso-occlusive crises. Current studies directed at perioperative management of sickle cell disease in various surgical specialties have continued to expand and modify the recommendations and options in caring for these surgical patients. These guidelines are important for the podiatric surgeon and the rest of the surgical team when planning surgery for a patient with SCD. A review of the podiatric surgical considerations when operating on a patient with SCD will be presented.

Since Keller and Beneson, and Caputi et al, there has been a relative paucity within the podiatric literature on the perioperative management of sickle cell disease in podiatric surgery.[2,3] In 1988, Smith et al reviewed the pathophysiology, diagnosis and lower extremity involvement in SCD.[4] This article seeks to provide the podiatric surgeon with an outline for preoperative, intraoperative, and postoperative considerations when podiatric surgery is planned for a sickle cell patient.

The goal in perioperative care of the sickle cell disease patient is to prevent red blood cell sickling. Conditions that can induce sickling include acidosis, dehydration, deoxygenation, low temperature/hypothermia, infection, and vascular stasis (Table 1). Anxiety and stress are secondary factors that can precipitate a sickle cell crisis and should be considered when caring for SCD patients. The surgeon, anesthesiologist, and the entire medical/surgical team must therefore direct their efforts towards prevention and correction of these conditions. Of course, in the event sickling does occur, treatment consisting primarily of hydration and narcotic analgesics should be initiated.

The cellular pathogenesis of sickle cell disease begins with the sickle-shaped erythrocyte that stems from the bone marrow with certain concentrations of HbS and total hemoglobin. These concentrations combined with the concentration of non-S hemoglobins (HA, HbA2, and HbF) reflect the potential amount of intracellular polymerization possible. Intracellular HbS polymerization occurs with erythrocyte deoxygenation (depolymerization may occur with reoxygenation unless the red blood cell has irreversible membrane damage from repeated polymerization cycles). Permanently polymerized erythrocytes include those with permanent membrane damage and dense cells with high mean corpuscular hemoglobin concentration. These cells with intracellular HbS polymerization lead to microvascular obstruction with resultant acute crisis and/or chronic, progressive organ damage, and are eventually eliminated by hemolysis and sequestration.[5] As a result, sickled red blood cells have a shorter life span, approximately 12-17 days versus approximately 120 days of a normal red blood cell. Thus, sickle cell disease patients often have hematocrits in the 20-25% range.[1]

Patients can exhibit different types of crises of variable severity and frequency depending on the extent of their disease. The vaso-occlusive crisis is the most characteristic type and often involves pain in the area of obstructed blood flow and a general fever. The severity of the crisis is primarily dependent on the vitalness of the tissue, for example, an occlusion to a cerebral artery resulting in a cerebrovascular accident may progress to death.[1]

It had previously been thought that in situ red blood cell sickling was the primary predetermining indicator of the clinical severity of sickle cell disease. However, current research has shown that the severity of the disease is more accurately defined

Table 1

PERIOPERATIVE MANAGEMENT OF FACTORS INVOLVED IN RED BLOOD CELL SICKLING*

Factor	Perioperative management	Pathophysiology of factors
Acidosis	•Maintain oxygenation and perfusion •Correct ketoacidosis with hydration and nutrition	•Aggregation occurs as HbS assumes deoxy conformation •Starvation ketoacidosis caused by poor nutrition •Reduced oxygen concentration in peripheral tissues
Dehydration	•Intravenous fluids (IVF) for at least 8 hours preoperatively •IVF postoperatively	•Increased blood viscosity and Hematocrit •Lower MCHC of HbS decreases risk of sickling •Vascular stasis secondary to dehydration •Inability to concentrate urine can lead to dehydration
Deoxygenation	•Preoperative blood transfusion to correct anemia and ensure oxygenation •Preoxygenate and use constant supplemental oxygen during and after surgery for 24 hours •Monitor oxygen saturation perioperatively •Incentive spirometry to prevent atelectasis postoperatively	•Oxygen tension < 40 mmHg causes HbS aggregation
Temperature changes/stress	•Keep patient warm and monitor their temperature (blankets, bair hugger, room temp) •Warm the IVF and any inspired gases	•Vasoconstriction and reduced oxygen to organs, can cause sickling/ishemia
Infection	•Prophylactic antibiotics (cover encapsulated organisms) •Treat aggressively and hydrate •Incentive spirometry/pulmonary toilet to prevent atelectasis, acute chest syndrome, and pneumonia	•Defective opsonization and macrophage phagocytosis •Alternative complement pathway •"Autosplenectomy" after age 6-8, at risk for encapsulated organisms •Vomiting/fever/chills can lead to dehydration •Mild leukocytosis with left shift is normal •Increases metabolic rate
Vascular Stasis	•Maintain high cardiac output and avoid hypotension •Intermittent compression pump for prolonged immobilization •Early postoperative ambulation	•HbS polymerization occurs with 2-4 minutes of stasis in hypoxic area

*Adapted from Lawrenz,[1] and Bassett et al[16]

by the extent and rate of intracellular HbS polymerization which in turn is determined by the amount of intracellular HbS composition and concentration as well as the percentage of oxygen saturation. Therefore, HbS polmerization is responsible for the red blood cell's rheological properties and their tendency to turn sickle-shaped.[5]

PREOPERATIVE CONSIDERATIONS

Every preoperative assessment should begin with a thorough history and physical. This includes identifying patients that may have undiagnosed or mismanaged sickle cell disease and referring them to a hematologist for further evaluation/treatment. For a known sickle cell patient, the podiatrist must remember that almost every organ in the body has the potential to be affected by this disease and therefore, preoperative management of the sickle cell patient starts with a complete history and physical with directed evaluation towards end-organ manifestations of the disease. To evaluate these possibilities, preoperative laboratory testing may include a complete blood count (CBC), coagulation studies, complete metabolic panel including liver enzymes, arterial blood gas, urinalysis, chest radiograph, electrocardiogram, and echocardiogram. Blood type and screening should be performed if the surgeon feels that a blood transfusion may be indicated. Hemoglobin electrophoresis quantitates the percentage of HbS present in the patient and may be a useful measure of the severity and relative perioperative risk potential for the individual patient.

A CBC of a sickle cell patient typically displays a normocytic, normochromic anemia with a decreased erythrocyte and increased reticulocyte (secondary to new red blood cell production) counts. Hemoglobin concentrations are often lower than the normal range and as a general rule levels below 7ηg/100 ml require postponement of any elective procedure. The hematocrit may range within 20-25%. Platelets are also often elevated. Increased blood breakdown products may be present in the urinalysis.[3]

Chest radiograph findings may include cardiomegaly, pulmonary congestion, or lung consolidations indicative of infarcts or pneumonia.[3] A baseline ABG and oxygen saturation will evaluate preoperative pulmonary function of the sickle cell patient, this will be important because these patients are more prone to pulmonary hypertension from chronic infarcts in the lungs.[6] Infarcts in the liver or kidneys may lead to hepatic or renal dysfunction. Therefore, liver enzyme studies and urinalysis are indicated. The electrocardiogram (EKG) of a sickle cell patient is often abnormal with sinus tachycardia, left axis deviation, and left ventricular hypertrophy as the most common findings.[3] These EKG changes are most likely a response to the tendency of sickle cell patients to be in chronic acidosis and hypoxia. It is also important to note that these patients will have an increased incidence of myocardial infarction and cerebrovascular accident perioperatively. It may therefore be necessary to attain a cardiology consult prior to and after surgery.

Chronic anemias can induce folic acid deficiency, thus folic acid supplements may be considered in the time period prior to surgery because decreased red blood cell production and circulation occurs with an aplastic crisis, which can occur secondary to a folic acid deficiency.[6] Prophylactic antibiotic therapy should be considered, especially since sickle cell patients have an increased infection rate compared to the general population. Preoperative narcotics and sedatives should be used with caution to avoid respiratory depression. Lastly, since the patient has been essentially fasting since midnight, preoperative intravenous fluids can decrease dehydration, hyperosmolality, and blood viscosity.[6]

Transfusion

Blood transfusions preoperatively seek to evade deoxygenation and to counter any existing anemia. It has been established that they lower blood viscosity, raise hemoglobin levels, and inhibit HbS production in the bone marrow. Decreasing the proportion of sickle red blood cells by transfusing packed red blood cells, can prevent oxygen tension levels below 40 mmHg, thereby avoiding HbS aggregation. Most studies agree that patients have a lower complication rate with a preoperative blood transfusion, but argue how aggressive the regimen should be prophylactically. A 1995 multicenter study performed by Vichinsky et al is often cited in the literature as convincing evidence that conservative regimens (Hb levels above 10 g per deciliter) are preferred over aggressive regimens (targeting HbS levels below 30%). This study concluded that the conservative regimen had significantly lower transfusion-associated complications while retaining equal effectiveness compared to the more aggressive regimen in prevention of perioperative complications.[7] The surgeon must bear in mind that a blood transfusion is by no means a benign event and the

benefits must outweigh the potential negatives. Complications associated with blood transfusions include alloimmunization, delayed transfusion reaction, transmission of viral infection/hepatitis, and iron overload.[8] Blood transfusions also deplete the immune system and therefore may predispose the transfused patient to an increased incidence of perioperative infection.

INTRAOPERATIVE CONSIDERATIONS

The intraoperative care of the SCD patient is primarily in the hands of the anesthesia team. Despite the choices the surgeon makes in terms of anesthesia and hemostasis, it will be the administration of appropriate intraoperative care to avoid the precipitating factors involved in red blood cell sickling that most affect the outcome of the patient.[9] These factors along with some preventive measures used perioperatively to counter them are listed in Table 1. They include prevention of acidosis, dehydration, deoxygenation, decreased temperature, infection, and vascular stasis.[1] Intraoperative diagnosis of a sickle cell crisis is challenging under general anesthesia, which often masks the clinical signs.[10]

Anesthesia

There are no studies directly evaluating the patient outcomes utilizing different anesthetic techniques in podiatric surgery. Regional, intravenous, and inhalation anesthesia have all been proven successful in SCD patients undergoing surgery.[6] Most authors suggest that the surgeon should continue to correlate the type of anesthetic they would ordinarily use with the procedure to be performed.[11] Most minor podiatric surgical procedures are performed with local anesthesia and monitored anesthesia care, and this continues to be an option for sickle cell patients. Hypotension is a potential risk of regional anesthesia and thus, intravenous fluids should be given if this method of anesthesia is chosen. Additionally, there is some evidence that there is compensatory vasoconstriction in unanesthetized areas with regional anesthesia.[11] The anesthesia team has an increased ability to control ventilation and therefore, oxygenation under general anesthesia. Whatever type of anesthesia is chosen, it is important to emphasize that the current literature reveals that the management of the anesthesia is much more important than the type of anesthetic used.[10]

The Cooperative Study of Sickle Cell Disease (CSSCD) is the most comprehensive study on patients with SCD. It was a 10 year multicenter study that recorded information of 3,765 sickle cell patients. Later, published literature analyzed certain subsections of the CSSCD data. One such study concentrating on surgical anesthesia included 1,079 surgical procedures (717 patients). The study results included: 1) postsurgical complications were more frequent in SCD patients who received regional anesthesia versus general anesthesia, and 2) perioperative transfusions were associated with a lower rate of SCD-related postoperative complications.[8] Information pertaining to podiatric surgical considerations can be extrapolated from this study, based on the author's criteria of distal extremity surgeries as low risk procedures. Postoperative complication rates were divided into non-SCD related complications and SCD-related complications and analyzed by type of anesthesia. For the low risk level, it was shown that non-SCD related complications (more commonly fever than infection and bleeding) were lower with general anesthesia compared with both regional or local anesthesia. SCD-related complications (most commonly painful crisis) were equally lower with general anesthesia and local anesthesia than with regional anesthesia.

Another study in the CSSCD series isolates the results gleaned from only the elective orthopedic surgery cases (118 patients, 138 surgeries).[12] Although these cases include foot and ankle surgeries that were considered low risk, the majority of the procedures were to the hip. The percentage of the orthopedic cases in the distal extremities were not specified, but by subtraction from given data they had to be of minimal quantity included in the study. Therefore, there is speculation as to whether the extensive data collected in these series of studies bears any conclusive evidence as to the amount of complications in distal extremity surgical procedures, and whether preoperative transfusion effects a lower complication rate when performing podiatric procedures on sickle cell patients. There have been no studies performed that isolate the distal lower extremity for analysis of complication rates. Of note, orthopedic cases of the low risk level (such as foot and ankle cases) were shown to have a statistically significant lowered rate of serious complications.[12] However, this again may not reflect any useful data for the podiatrist given the ability for the predominance of hip surgeries to skew the complication data based more on the intrinsic complication rate of the type of hip surgery more than the risk level (22 out of 58 low risk procedures were hip