Major Surgery in a Jehovah's Witness with Sickle Cell Disease

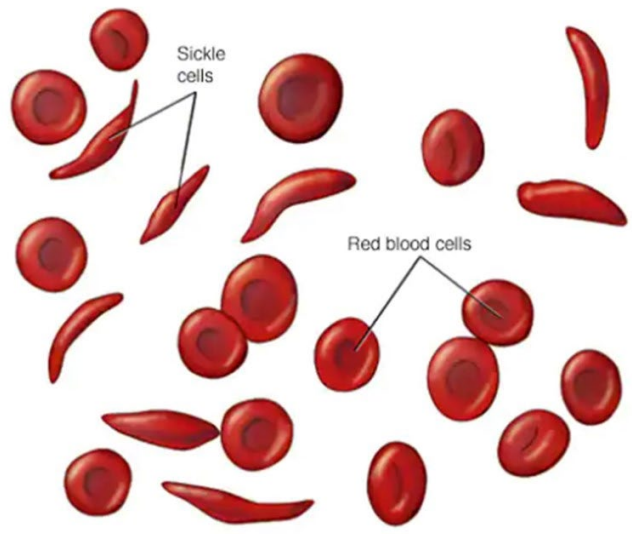

Photo courtesy of Mayo Foundation for Medical Education and Research

Introduction

Major surgery is often associated with high intraoperative blood loss and blood transfusion. Blood transfusion has hitherto been the default therapy for sickle cell disease (SCD) patients. Jehovah's Witnesses (JW's) decline blood transfusion for religious reasons.

Major surgery, elective or emergency, can be performed safely in JW's by employing bloodless surgery techniques perioperatively. In JW's with SCD, additional measures and precautions are needed to prevent complications due to sickling.

Sickle Cell Disease

SCD is a group of haemoglobinopathies characterized by sickling of red blood cells, chronic hemolytic anemia, painful vaso-occlusive crises, and acute and chronic end-organ damage. First described by James Herrick in 1910, it is the commonest inherited blood disorder, and affects > 30 million people worldwide, mainly persons of sub-Saharan descent, but also persons from South Asia, the Middle East, and the Mediterranean.

Normal haemoglobin is HbAA with 2 α and 2 β globin chains. The abnormal haemoglobin in SCD, HbS, on deoxygenation is prone to polymerization, causing sickling, rigidity of RBC, hemolysis, and vaso-occlusion. HbS causes disease only when both β globin chains are abnormal (HbSS, HbSβ, HbSC, etc). HbAS is thus sickle cell trait, not considered a disease.

Factors which precipitate sickling in SCD are hypoxia, dehydration, acidosis, hypothermia, hypotension, pain, hyperkalemia, increased blood viscosity, vascular stasis, vasoconstriction, infections (bacterial, viral, or protozoal), extremes of weather, and stress.

The Challenge of Surgery in Sickle Cell Disease

SCD patients may require surgery due to complications of SCD, e.g., hip replacement for necrosis of femoral head, cholecystectomy for gall stones, and splenectomy for hypersplenism. On the other hand, surgery may be incidental in SCD patients, e.g., caesarean section, cardiac surgery, and appendectomy.

Surgical procedures in SCD are associated with increased risks of peri-operative mortality and morbidity from vaso-occlusive (painful) crisis, acute chest syndrome, acute kidney injury, cerebrovascular accident, congestive heart failure, post-operative infections, and venous thromboembolism. Majority of complications occur postoperatively.

Problems of transfusion in SCD

Routine preoperative transfusion in SCD patients is now discouraged due to hazard of alloimmunization, hemolytic and non-hemolytic reactions, hyperhaemolysis, infections, and increased blood viscosity which can result in sickling. Other hazards of blood transfusion such as hyperkalemia, hypothermia, acidosis, and hypoxia due to storage lesions can also induce sickling in SCD patients.

Pillars of Bloodless Surgery

Bloodless surgery techniques are grouped under 4 'pillars': 1) Optimizing the hematocrit (HCT); 2) Minimizing blood loss; 3) Optimizing tissue oxygenation; and 4) Supporting patient's tolerance of anemia. All the techniques under the 4 pillars may find application throughout the perioperative period for all patients including SCD patients, but they are selected according to what is appropriate for and acceptable to the individual patient.

Prevention of complications of surgery in SCD

Prevention of complications of surgery in SCD requires meticulous perioperative care, which involves full preoperative assessment, adequate hydration, thermoregulation, avoidance of hypoxia, adequate pain control, and indeed, avoidance of all factors predisposing to sickling as outlined earlier.

Preoperative Care

Preoperative Assessment. History and physical examination of SCD patient will include ascertaining the frequency of crisis and the date of the patient's last crisis, known triggers for crisis, baseline level of activity, baseline opioid use, steady-state hemoglobin and hematocrit, assessing for co-morbidities, and cardiac and pulmonary complications.

Investigations will include complete blood count, iron studies, genotype confirmation, baseline oxygen saturation (SpO_2 by pulse oximetry), urinalysis, serum urea, electrolytes, and creatinine, chest x-ray, and other investigations as indicated,

e.g., ABG, Pulmonary Function Tests, ECG, LFT, and neurological imaging).

A multidisciplinary review of the patient is necessary, and would involve at least the surgeon, anesthesiologist, hematologist, pain management team, nutritionist, and chest physician.

Optimizing the Hematocrit Preoperatively. Target HCT in SCD preoperatively should ≥ 30% (Hb ≥ 10 g/dL). The hemoglobin may be successfully raised with folic acid 5mg/day, Vitamin B12 150 µg/day, Vitamin C 500 mg/day, nutritional support (green vegetables, fruits, proteins, and water), erythropoietin 100-150 U/kg s.c. on alternate days, and cautious iron therapy.

Iron overload in SCD is related to multiple transfusions. Iron deficiency anemia has been found to be a significant problem in young non-transfused SCD patients, and it responds to iron therapy. Pregnant sicklers who have *not* been transfused are also predisposed to iron deficiency anemia due to urinary losses from intravascular hemolysis and increased dietary requirement in pregnancy, so they can benefit from iron supplementation. Serum ferritin < 25 ng/mL and low MCV are useful screening tools.

Other Preoperative Measures and Intervention. The patient would need to be admitted ahead of surgery, and a cold environment should be avoided. Phlebotomies should be restricted, to avoid anemia.

Adequate hydration should be ensured, and hypotonic fluids are preferred (4.3% dextrose in 1/5 saline). Much normal saline can result in acidosis and sickling, and excessive intravenous fluids can result in pulmonary edema, which predisposes to acute chest syndrome. Prolonged preoperative fast should be avoided.

Anticoagulants, NSAIDs, and herbal supplements can lead to increased hemorrhage, and should be stopped days ahead of surgery. Hydroxyureas, L-Glutamine, and other newer drugs can help prevent or mitigate crisis.

The patient's choices of bloodless surgery techniques should be ascertained, written informed consent obtained, and any objections documented. The Hospital Liaison Committee of Jehovah's Witnesses can be helpful in the holistic care of the JW patient.

Intraoperative Care

Normothermia is important not only to avoid sickling but to minimize blood loss. Operating room temperature should be maintained at ≥ 27° C. Thermal suits or blankets may be used as appropriate, and intravenous infusions should be warmed if necessary.

Anesthesia. Spinal or epidural anesthesia is preferred for less blood loss and better pain control postop with epidural. However, there is need to beware of hypotension and pooling of blood which can result in sickling. Adrenaline should be avoided, and dopamine may be used if inotropic agent is needed.

Tissue oxygenation should be optimized through pre-oxygenation and maintenance of SpO_2 above 96% or basal level, whichever is higher, using 30-50% inspired oxygen (Henderson 1994).

Meticulous volume replacement is important, but caution should be exercised with normal saline, and fluid overload should be avoided for the reasons stated earlier. Colloids should be used for blood loss ≥ 1000 mL or ¼ of the patient's blood volume. The patient's blood pressure should be maintained just above the lower end of normal to minimize blood loss.

Monitoring. Continuous oxygen monitoring (pulse oximetry) is an important component of the standard monitoring during anesthesia in SCD patients. Arterial blood gases are sometimes necessary to confirm hypoxia. Urine output should be monitored by urethral catheterization.

Bloodless surgery techniques. The patient should be positioned with the op site above the right atrium and avoiding pressure on large vessels (e.g. pregnant women are turned slightly to the left) to reduce blood loss. Pharmacological agents to minimize blood loss should also be used pre-emptively whenever possible, e.g., tranexamic acid, vitamin K, Epsilon aminocaproic acid, Desmopressin, and recombinant Factor VIIa.

Meticulous hemostasis is important, and a dry field should be maintained by pre-emptive and prompt ligation of vessels. Judicious use of topical hemostats, electrosurgery, harmonic scalpel, CUSA knife, laser, plasma jet, and other devices, can help minimize blood loss. Laparoscopic and robotic surgery are also associated with less blood loss.

Blood salvage/cell salvage can be used safely in SCD, as can tourniquet. Use of acute normovolemic hemodilution has been reported in sicklers but seems to be rarely needed nowadays.

Postoperative Care

Postoperatively, oxygen therapy should be continued to maintain SpO_2 above 96% or basal level, whichever is higher, for 24 hours to support the patient's tolerance of anemia. Continuous oxygen monitoring should be maintained until SpO_2 is sustained at baseline with room air.

Adequate analgesia should be administered to avoid hypoxia which can result in sickling. Normal haemodynamic status and fluid balance should be maintained to avoid hypotension, dehydration, and fluid overload. Fluid administration should not exceed 1½ times patient's maintenance fluid requirement to avoid pulmonary oedema which can result in acute chest syndrome as stated earlier. Incentive spirometry helps to prevent atelectasis and acute chest syndrome.

Prompt treatment of febrile illness, infection, or sepsis should be done to avoid sickling. Anaemia should be treated according to severity with iron therapy, erythropoietin and adjuncts outlined earlier.

Outcomes of Bloodless Surgery in SCD patients

A systematic review and meta-analysis of randomized and observational studies by Alotaibi and co-workers (2014) comparing transfusion vs non-transfusion strategy in SCD found no difference in perioperative mortality, and vascular and non-vascular complications. Steven Frank, MD, (Medical Director: Bloodless Medicine and Surgery Program, Johns Hopkins Hospital, Baltimore, USA) based on his own studies in a wide spectrum of patients stated: “the use of appropriate blood conservation measures for patients who do not accept transfusions results in similar or better outcomes.”

Conclusion

Major surgery in SCD patients is associated with higher morbidity and mortality than in the general population. Blood transfusion in SCD is associated with alloimmunization, haemolytic & non-haemolytic reactions, hyperhaemolysis, infections, and increased viscosity predisposing to sickling.

Major surgery can be performed safely in JW’s with SCD by judiciously employing bloodless surgery techniques with good outcomes. Experience gained in bloodless surgery with JW’s can benefit non-JW SCD patients too!

Contributor: Nathaniel Usoro, MD, FWACS, FICS

Further Reading

1. Sedrak A, Kondamudi NP. Sickle Cell Disease. In: *StatPearls*. StatPearls Publishing; 2021. Accessed December 7, 2021. http://www.ncbi.nlm.nih.gov/books/NBK482384/

2. Walker I, Trompeter S, Howard J, et al. Guideline on the peri-operative management of patients with sickle cell disease. *Anaesthesia*. 2021;76(6):805-817. doi:10.1111/anae.15349

3. Gohel MS, Bulbulia RA, Slim FJ, Poskitt KR, Whyman MR. How to approach major surgery where patients refuse blood transfusion (including Jehovah’s Witnesses). *Ann R Coll Surg Engl*. 2005;87(1):3-14. doi:10.1308/1478708051414

4. Usoro NI. Bloodless Medicine and Surgery. In: *Blood Transfusion in Clinical Practice*. IntechOpen; 2012:191-201. https://www.intechopen.com/books/blood-transfusion-in-clinical-practice/bloodless-medicine-and-surgery

5. Vichinsky E, Kleman K, Embury S, Lubin B. The diagnosis of iron deficiency anemia in sickle cell disease. *Blood*. 1981;58(5):963-968.

6. Iron supplementation in pregnant sicklers: an opinion | BMC Pregnancy and Childbirth | Full Text. Accessed December 8, 2021. https://bmcpregnancychildbirth.biomedcentral.com/articles/10.1186/s12884-018-1894-y

7. Alotaibi GS, Alsaleh K, Bolster L, Sean McMurtry M, Wu C. Preoperative transfusion in patients with sickle cell disease to prevent perioperative complications: A systematic review and meta-analysis. *Hematology*. 2014;19(8):463-471. doi:10.1179/1607845414Y.0000000158

8. Helwick C. Conserving Blood During Surgery Safer Than Transfusion. Medscape Conference News (ASA 2014). Published October 22, 2014. https://www.medscape.com/viewarticle/833649