Anesthetic Considerations for Patients with Sickle Cell Disease

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Introduction

Sickle cell disease (SCD) is an autosomal recessive disease prevalent in African, Middle Eastern, and Central Indian populations as it confers immunity against malaria.[1,5] Healthy red blood cells (RBCs) have a half-life of ~120 days, however this is reduced to 10-12 days in SCD causing chronic hemolytic anemia, hyper-proliferative bone marrow and hyperdynamic circulation, affecting all organ systems. Deoxygenation can cause a cascade of events leading to a sickle cell crisis. Some common triggers are pain, dehydration, and cold temperatures.[1]

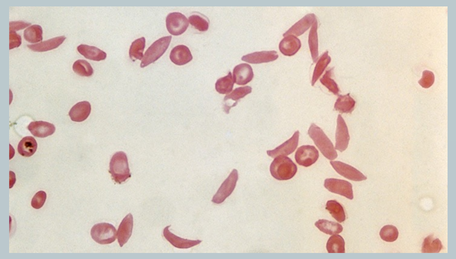

Figure 1. Microscope image of sickled RBCS

Objectives

- Discuss challenges in intraoperative management of SCD patients
- Develop pain management approaches for postoperative pain and SCD crises
- Compare the benefits of general and regional anesthesia
- Evaluate the risks associated with different transfusion regimens

Review of Literature

Preoperative Care

A thorough preoperative evaluation is imperative for SCD patients and is aimed at identifying baseline Hgb and transfusion requirements, organ dysfunction, preventing crises, and postoperative complications. SCD patients are prone to cardiac and pulmonary complications and dysfunction, which may necessitate a TEE/ECHO. Difficult IV access due to chronic damage to endothelial lining, repeat blood draws and placement of catheters is common.[2] Incentive spirometry can decrease the incidence of atelectasis and ACS.[1]

Fluid Management

It is imperative that patients are adequately hydrated. IV hydration at the standard maintenance rate is sufficient as fluid overload may lead to acute chest syndrome (ACS). NS is acidic and may increase blood viscosity and cause acidosis.[1,2] A pediatric study found NS bolused patients had poorer pain control compared to with other types of fluid. Isotonic fluids have not been thoroughly studies.[2] At this time, the official recommendation is still hypotonic fluid.

Intraoperative Management

Goals: avoid hypoxemia, hypercapnia, acidosis, hypothermia, and hypovolemia. Oxygen supplementation is not usually necessary as oxygen toxicity is known to suppress the formation of new RBCs and cause lung injury. ABGS may provide more accurate information regarding oxygen saturation as pulse oximetry readings may not reflect arterial oxygen tension.[1,5]

Regional Anesthesia

The benefits of RA are unclear. Research shows SCD complications are more frequent in patients receiving RA, however, these were sicker patients. RA theoretically causes regional hypoperfusion, venous stasis, redistribution of blood flow, oxygen tension and vasoconstriction of non-blocked areas.[1] It may aid in pain control, reduce length of stay, and decrease opioid consumption. It is recommended for local pain not adequately treated with opioids. Nerve catheters do not show decrease in reported pain scores.[3] Use of tourniquets are controversial because they create ideal conditions for sickling.[1]

Review of Literature cont.

Pain Control

Pain is treated aggressively and there is a large emphasis on multimodal pain management.[5] This includes opioids, NSAIDs, and ketamine. Ketamine may be useful for pain refractory to opioid therapy, but will not reduce for more than 3 days after initiation of therapy and does not decrease opioid consumption.[4] Opioid PCAs should be used with caution. Nonpharmacologic measures include music, heat and ice packs.[1]

Transfusion Protocols

Preoperative blood transfusion is controversial. Transfusion can reduce HbS% and improving oxygen delivery, however, it will increase blood viscosity and the risk of alloimmunization. Net benefit if Hb ≤ 10 g/dL.[1] During acute events, transfusion may not be required in patients if Hb > 9 and no hypoxemia.[5] Aggressive transfusion regimens aim for Hb ≥ 10 g/dL AND HbS% < 30%. Conservative regimens do not have an HbS% goal. There is no difference in outcomes or blood loss in aggressive vs conservative transfusion regimens, however the aggressive regimen had increased report of alloimmunization.[2,6] Patients should receive preoperative transfusion as another study in which patients did not receive preoperative transfusions was terminated due to poor outcomes and increased risk of VOC and ACS. All patients undergoing GA > 1 hour should receive transfusions. Simple transfusions are recommended for Hb < 8.5 g/dL, with posttransfusion Hb goals < 11 g/dL. Exchange transfusions should occur if Hb > 8.5 g/dL. Autologous cell salvage has reported varying degrees of success and is not currently recommended.[5]

Acute Chest Syndrome (ACS)

ACS is an acute lung injury characterized by respiratory symptoms, fever, and new radiodensity on chest imaging. Patients are at risk of ACS in the immediate post-operative phase and is the leading cause of death in patients presenting to the ED. Adults commonly present with chest pain, SOB, and neurologic findings. Pediatric patients present with fever and wheezing. VOC is the most common inciting cause along with respiratory illness.[5] This risk increases with the use of opioids and excessive IV fluids.[5]

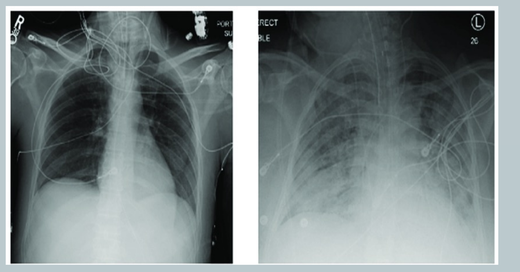

Figure 2. Chest x-ray of healthy lungs (left). Chest x-ray of acute chest syndrome (right).

Patients should be treated with adequate ventilation, transfusion, antibiotics, and bronchodilators.[1] ACS can cause worsening lung function due to fibrosis, and overtime can increase PHTN. Intercostal nerve blocks are recommended for patients with ACS and pain not adequately treated with opioids. Up to 21% of patients will experience respiratory failure within 24 hours of symptoms.[5]

Conclusions

Most of these studies were conducted on otherwise healthy and optimized patients and undergoing elective procedures of low to moderate risk. The benefits of GA vs RA need more research as it is unclear if RA is actually beneficial. It was difficult to find reports regarding management of patients undergoing emergent procedures. More research is needed on different pain management techniques for patients undergoing SCD crises and optimal fluid choice for volume resuscitation.

References

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