Perioperative management of children with sickle cell disease undergoing major abdominal surgery

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Sickle cell disease (SCD), which is due to homozygous inheritance of the hemoglobin S (HbS) variant is one of the commonly inherited hemoglobinopathies in the Eastern Province of the Kingdom of Saudi Arabia (KSA). Affected children often require operative interventions to treat SCD related complications, but for many years, surgery on SCD children was associated with an unacceptably high complication rate, sometimes as high as 50%.[1] This high complication rate is mostly attributed to intraoperative events such as hypoxia, hypovolemia, hypothermia and acidosis. In an attempt to obviate or minimize this, a variety of perioperative management protocols including preoperative blood transfusion (BT) regimens have been adopted.[2-6] In an attempt to decrease the perioperative complications of children with SCD undergoing major abdominal surgery we adopted a perioperative management protocol. The protocol consists of: (1) Thorough preoperative clinical evaluation (2) Complete blood count on admission, preoperatively, and postoperatively, (3) Preoperative BT for those with an admission Hb<10gldl. This was carried out with simple BT using packed red blood cell (RBC) and the aim was to increase the hemoglobin (Hb) to 10-12 gldl and the hematocrit to 30-40%. The amount of blood to be transfused was calculated roughly using the following formula: (Hb desired-Hb on admission) x 3 x weight. No attempt was made to estimate the levels of HbS and hemoglobin F (HbF) preoperatively. (4) Preoperative hydration with intravenous fluids starting the night before operation or immediately at the time of admission for emergency operation. The intravenous fluids were given at the maintenance requirements of 1.5 and depending on the age of the patient either 5% dextrose in .5 normal saline or 5% dextrose in .25 normal saline was given. The intravenous fluids were continued postoperatively until the patient was able to take enough fluids orally.[5] Intraoperatively, hypoxia, hypothermia and acidosis were avoided and oxygenation was continuously monitored by pulse oximetry.[6] Postoperatively, the patients received analgesia in the form of pethedine 1mg/kg intramuscular (IM) every 4-6 hours regularly for the first 24 hours and then as required.[7] Early ambulation was encouraged postoperatively, as well as chest physiotherapy using incentive spirometry. This study is an evaluation of this protocol in 25 consecutive children with SCD who had major abdominal surgery during a 2 year period from November 1998 to November 2000. There were 16 males and 9 females. Their ages ranged from 5-12 years (mean 8.4 years). Their mean HbS level was 74% (57.8-91.8%), and their mean HbF level was 24.9% (5.1-41%). The different types of operations performed are shown in **Table 1**. Nine patients had splenectomy and in all the indication for splenectomy was recurrent splenic sequestration crisis. Splenectomy was carried out through a left upper abdominal transverse incision. Six patients had splenectomy and cholecystectomy. One of them had a splenectomy due to hypersplenism while the other 5 had splenectomy for recurrent splenic sequestration crisis. In these 6 patients, cholecystectomy was carried out for asymptomatic gallstones discovered incidentally by ultrasound, and both splenectomy and cholecystectomy were carried out through a left upper abdominal transverse incision. One patient had splenectomy, cholecystectomy and repair of paraumbilical hernia through a left upper abdominal transverse incision. Splenectomy in this patient was due to recurrent splenic sequestration crisis, and cholecystectomy for asymptomatic gallstones. Eight patients had cholecystectomy: 3 had laparoscopic and 5 had open cholecystectomy. Cholecystectomy was carried out for symptomatic gallstones in one patient. Another one patient had open cholecystectomy and appendecectomy, while another had open cholecystectomy and excision of right

Table 1 - Types of operations performed.

Type of operation	n of patients
Splenectomy	9
Splenectomy and cholecystectomy	6
Open cholecystectomy	3
Laparoscopic cholecystectomy	3
Appendicectomy for perforated acute appendicitis	1
Open cholecystectomy and appendicectomy	1
Splenectomy and cholecystectomy and repair of paraumbilical hernia	1
Open cholecystectomy and excision of right ovarian dermoid	1

n - number

ovarian dermoid cyst. Both were carried out through a right upper abdominal transverse incision. Interestingly in all those who had cholecystectomy whether for symptomatic or asymptomatic gallstones, there was histological evidence of chronic cholecystitis. One patient had appendecectomy for perforated appendix. The mean operative time was 72.4 minutes (range 35 minutes in one patient who had splenectomy and120 minutes in one patient who had splenectomy and cholecystectomy). Their hematological parameters are shown in **Table 2**. Two patients admitted with a Hb of 9.8 gldl and 10.7 gldl respectively did not receive preoperative BT. Three patients were admitted with major splenic sequestration crisis with an admission Hb of 3.8, 4.1 and 4.3 gldl. They were transfused preoperatively with 1700 cc, 1050 cc, and 800 cc of packed RBC respectively. The mean preoperative BT was 441.7 cc of packed RBC (200-1700 cc). None of our patients required postoperative or intraoperative BT, and in all except 4, the postoperative Hb was higher than the preoperative value. There was no mortality. None of our patients developed postoperative vasoocclusive crisis, and only one patient developed a mild acute chest syndrome (4%). This was following appendesectomy for a perforated appendix. This patient was admitted with a Hb of 10.7 g/dl and was not given BT. The mean hospital stay was 7.4 days (3-20 days) while the mean postoperative stay was 5.3 days (3-9 days). Children with SCD undergoing surgical procedures under general anesthesia are at increased risk of perioperative complications, particularly vasoocclusive crisis and acute chest syndrome. These complications are part of the disease process, and usually secondary to intraoperative events such as hypoxia, hypovolemia, hypothermia and acidosis. Although these complications can be seen following any operative procedure, they are more commonly seen in those undergoing thoracotomy or laparotomy.[6] Earlier studies reported perioperative mortality rate as high as 10% and a postoperative complication rate up to 50%,[1] and due to this unacceptable high complication rate, surgery was not advocated except in symptomatic patients. Nowadays and inspite of better understanding of SCD, improved anesthetic and surgical techniques, and careful perioperative management, the perioperative complication rate in these patients is still high. To minimize these risks, a variety of perioperative management protocols, including different preoperative BT regimens have been adopted. Perioperative BT to correct anemia and decrease the percentage of HbS level is beneficial in reducing the rate of SCD related complications, but the amount of BT necessary for safe surgery is not known. Adams et al[2] reviewed 92 children with SCD who had 130 surgical procedures, and received an aggressive BT regimen to alleviate anemia and reduce HbS level to less than or equal to 30% and 10% major postoperative complications was reported. In a multicenter trial comparing a conservative BT regimen to increase the Hb level to 10 gldl to an aggressive BT regimen designed to lower the HbS level to less than 30% in a total of 551 patients who underwent 604 operations, found the frequency of serious postoperative complications similar in the 2 groups, namely 35% in group 1 versus 31% in group 2. Furthermore, the conservative transfusion regimen resulted in only half as many transfusion-related complications. They reported a mortality rate of 0.3%, and among the serious complications, acute chest syndrome occurred in approximately 10% of each group.[4] Griffin and Buchanan[6] reviewed 54 children with SCD who had 66 operative procedures without preoperative BT and reported a 26% overall postoperative complication rate, but the complication rate increased to 50% in those undergoing major surgery. Ware et al[5] reviewed 27 children with SCD who had elective cholecystectomy and received preoperative BT regimen to achieve a Hb concentration of 11-14 gldl with greater than 65% HbA and no reported perioperative morbidity or mortality. Bhattacharyya et al[3] reviewed 22 children with SCD who underwent cholecystectomy and received preoperative BT to achieve a Hb level of

Table 2 - Hematological parameters.

Parameters	Admission Hb (gldl)	Admission HCT %	Preoperative blood transfusion	Preoperative Hb (gldl)	Preoperative HCT %	Postoperative Hb (gldl)	Postoperative HCT %
Mean	7.2	21.1	441.7cc	10.5	31	11.8	11.8
Range	3.8 - 10.7	9.7 - 34.4	200 - 1700	9.5 - 12.3	26.3 - 34.8	9.5 - 13.8	9.5 - 13.8

Hb - hemoglobin, HCT - hematocrit

more than 9 gldl and HbS level less than 30% was reported on SCD related vasoocclusive complications.[3] In our study, the simple BT preoperatively was as effective as the traditionally aggressive transfusion therapy or exchange transfusion therapy in reducing postoperative complications, even in children undergoing major abdominal surgery. The mean preoperative BT in our patients was 441.7 cc of packed RBC. This is of importance in our setting where BT is not readily available, and inspite of careful blood donors screening, the risk of transmission of blood born infections remains a concern in multiple-transfused patients. Furthermore, the risk of allosensitization, which is reported in 15-20% of SCD patients, increases with increase in transfusion therapy.[7] Acute chest syndrome (ACS) is a well documented postoperative complication in patients with SCD with an incidence ranging from 0.4-10%.[4] Only one of our patients (4%) developed postoperative ACS, and in this patient ACS was mild. This low incidence in our patients may partly be due to the high HbF level (mean HbF level was 25%), which has a protective effect and reduces the risk of postoperative ACS.

In conclusion, preoperative BT may not be indicated for children with SCD undergoing minor surgery under general anesthesia, it is indicated in those undergoing major surgery. Simple BT is as effective as an aggressive transfusion regimen, and even in children undergoing major abdominal surgery. The importance of maintaining an adequate hydration, and preventing stimuli for sickling such as hypoxia, hypothermia, acidosis and stasis cannot be over emphasized.

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Erratum

In manuscript “Clinical and prognostic values of anti-hepatitis B core immunoglobulin M detection in asymptomatic hepatitis B surface antigen carriers” Saudi Medical Journal 2002; Vol. 23 (7): 777-781, the title of Table 1 should have appeared as follows: Possible risk factors for acquisition of HBV in HbsAg/Anti-Hbe carriers with or without anti-HBc IgM.

Erratum

In manuscript “Pyoderma among Hajj Pilgrims in Makkah” Saudi Medical Journal 2002; Vol. 23 (7): 782-785, the spelling of this drug should have appeared as follows: cotrimoxazole.

Erratum

In manuscript “Rapidly growing mycobacterial pulmonary infection in association with severe gastroesophageal reflux disease” Saudi Medical Journal 2002; Vol. 23 (7): 854-856, the date of received should have appeared as follows: Received 21st January 2002.