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CASE REPORT

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Can children with sickle cell disease undergo open splenectomy without preoperative transfusion despite severe anemia? A report of three cases

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Abstract Preoperative red cells transfusion to correct anaemia and to reduce the proportion of sickle red cells is part of standard preparation of children with sickle cell anaemia (HbSS) for major procedures including open abdominal surgeries. We report three children with sickle cell anaemia presenting with chronic massive splenomegaly and hypersplenism. The children were initially denied surgery because of extremely low haemoglobin levels and the inefficacy of transfusion. Subsequently, they underwent successful open abdominal splenectomy without any red cells transfusion. These observations are important to paediatricians and surgeons in settings where HbSS is common. They highlight the fact that surgery should not be withheld from children with sickle cell anaemia and massive splenomegaly purely on the basis of difficulty in correcting anaemia before the procedure.

Key words: Sickle cell disease, surgery, splenectomy, transfusion.

Introduction

Red cells transfusion to reduce the proportion of sickle erythrocytes and to correct anaemia is part of standard preoperative management of children with sickle cell anaemia (HbSS), undergoing moderate or high-risk procedures such as open abdominal surgeries.[1-3] However in an Africa setting, applying this preoperative care may be difficult due to the unavailability of safe blood products or certain clinical conditions that may render transfusion impossible. This may result in an anaesthetist or surgeon denying the patient surgical procedures. Such a situation poses a serious concern to the paediatrician because of the predictably poor short-term outcome of splenomegaly in HbSS disease. Here we report three children with HbSS who successfully underwent open splenectomy without any preoperative red cell transfusion, despite the very low values of their haemoglobin, demonstrating that it is possible for the paediatrician to overcome this dilemma.

Case presentation

Patient A, born on April 29th 2006 was diagnosed HbSS at the age of nine months and enrolled at the age of 12 months into our established comprehensive clinical care programme (CCCP), which includes an intensive socio-medical intervention programme to ameliorate the disease course in severely ill children.[4] On admission he presented with jaundice, enlarged spleen two centimetres below the left costal margin, had a weight of 11,220 grams and a height of 76 cm.

During the first 24 months of follow-up, several SCD-related acute events were registered. These included three mild vaso-occlusive crises successfully managed on outpatient basis with aspirin and acetaminophen; three episodes of fever without focus, two of which required hospitalization and intravenous antibiotics; and notably the initiation of hypersplenism with progressive enlargement of spleen, reaching the left iliac fossa. This was associated with a progressive decrease in the values of haemoglobin, declining from 65 g/L to 39 g/L. Two attempts at red cell transfusion to correct the anaemia resulted in acute abdominal pain associated with an increase of spleen volume with no substantial change of value of haemoglobin level. Splenectomy was proposed, but both paediatric surgeon and anaesthetist dissented because of his very low haemoglobin level, the inefficacy of red cell transfusion and the fear of peri-operative complications leading to death.

However, several discussions resulted in an agreement on the following protocol, which was approved by the Faculty of Health Sciences Ethics Committee. (1) In addition to administration of a booster injection of 23 valence polysaccharide pneumococcal and meningococcal vaccines three weeks before the procedure, the preoperative preparation would comprise hydration with intravenous fluids (6-7 ml/kg/hour), starting 24 hours before the procedure, administration of oxygen, prophylactic intravenous administration of bactericidal antibiot-

ics, as well as careful monitoring of serum electrolytes, heart rate, pulse oximetry and urinary outflow. (2) The intra-operative management would comprise monitoring of respiratory and heart rates, pulse oximetry, blood pressure, electrocardiographic features and haemoglobin level; in addition, appropriate bleeding control to minimize intra-operative blood loss would be combined with red cells transfusion after the ligature of the main splenic artery. (3) The post-operative management would comprise continuation of the intravenous hydration and antibiotics, quinine administration until the third post-operative day, at least, intravenous acetaminophen and codeine to control postoperative pain, and recording of all peri-operative clinical events.

After obtaining informed written parental consent, the child underwent open abdominal surgery (laparoscopic equipment was not available) on April 8th 2009, at the age of 36 months under general anaesthesia. The procedure lasted 75 minutes and the surgeon performed partial splenectomy, removing 850g portion of spleen; the retained portion represented approximately 20% of the spleen. Because no significant intra-operative changes were observed in heart rate, pulse oximetry, blood pressure, electrocardiographic features and values of haemoglobin level, it was decided to refrain from the planned intra-operative red cells transfusion. The recovery room stay lasted three days and no adverse clinical events were recorded post-operatively. Progressive increase in haemoglobin levels were observed, reaching a steady state level of 7.5 ± 5 g/L up to six months post-operation, without any red cell transfusion. A transient thrombocytosis was observed for which patient received daily aspirin at 10 mg/kg for 45 days.

Given these encouraging results, two other children, with a similar clinical presentation, previously denied surgery, were then accepted for open abdominal splenectomy. Patient B was born on January 29th 1995 and Patient C was born on November 4th 2004. They were enrolled into the CCCP on September 20th 2001 and March 15th 2006 respectively and underwent the procedure on June 10th 2009 (at 172 months) and on September 16th 2010 respectively (at 70 months). About 90% of the spleen (1,200g) was removed in patient two while about 85% (1,100g) was resected in patient three. Peri-operative fever without any identified focus of infection was recorded in patient two on the first post-operative day. A third generation cephalosporin, Ceftriaxone was added to treatment. In patient three, cellulitis of the left leg was observed on day 34 post-operatively, requiring a two day hospitalization and intravenous antibiotics.

Additionally, two episodes of vaso-occlusive crises were recorded post-operatively on days 118 and 140, respectively. None of these two patients required red cells blood transfusion. Patients' demographic and pre-operative clinical characteristics are detailed in Table 1 and the post-operative growth, platelet count and haemoglobin levels are given in Table 2, Progressive increase of steady-state haemoglobin levels was observed reaching 78 ± 3 g/L and 81 ± 2 g/L in patients 2 and 3, respectively up to 6 months post-operation. All patients showed a physical growth recovery pattern: during the first 12 months post-operation, the weight and height increases were 4,500g, 5,500g, 4,840g and 13cm, 7cm, 9cm for patients one, two and three, respectively, compared to 2,000g, 1200g, 1400g, respectively, during the follow-up time before surgery. At the time of this report, no further long term adverse event was recorded in any of the patients. Abdominal ultrasound scan performed 50, 48 and 34 months post-operatively in patients one, two and three, respectively, showed no re-growth of the residual spleen.

Table 1: Patient Demographics and Pre-operative Clinical Characteristics

Characteristic	Patient A	Patient B	Patient C
Age (months)	36	172	70
Sex	Male	Male	Male
Weight (grams)	15,000	20,500	14,900
Height (cm)	94	126	109
Length of follow (months)	24	93	54
SCD history			
Painful crises	3	2	2
Acute chest syndrome	0	0	0
Fever without focus	3	2	5
Malaria attack	0	2	1
Focal infection	0	3	2
Transfusion	2	4	0**
Hospitalization	3	2	0
Other medical history	None	None	None
Weight increase* (grams)	2,000	1,200	1,400
Height increase* (cm)	10	3	10
Hemoglobin level (grams/L)	39	37	31
Platelets count (x 10^9/L)	59	88	148

* Observed increase during the follow-up before surgery
** This patient had never received a blood transfusion

Table 2: Patients Post-operative Data on Growth, Platelets Counts and Hemoglobin Levels

Characteristic	Patient A	Patient B	Patient C
Clinical events	None	None	3
Weight (grams)			
Month + 1	14,000	17,500	14,200
Month + 3	16,000	20,000	16,420
Month + 6	17,200	22,000	17,880
Month + 12	18,500	23,000	18,840
Height (cm)			
Month + 1	95	126	110
Month + 3	98	127	113
Month + 6	103	131	118
Month + 12	108	133	119
Hemoglobin level (grams/L)			
Month + 1	55	51	60
Month + 3	65	67	77
Month + 6	75	76	82
Month + 12	78	80	80
Platelets count			
Month + 1	857	981	704
Month + 3	650	631	585
Month + 6	450	535	486
Month + 12	330	440	420

Discussion

In children with HbSS, major surgery without preoperative transfusion, has been reported to be associated with serious peri-operative complications.[1] Therefore, these children usually receive pre-operative red cell transfusion. However, the cases presented herein demonstrate that in desperate situations, close cooperation among the paediatrician, anaesthetist, and surgical team can still permit the procedure without preoperative transfusion and with minimal risk of peri-operative-related mortality and morbidity. Despite the very low pre-operative haemoglobin levels and the open abdominal surgery, no serious peri-operative or post-operative complications were recorded in the three cases. Some recent studies recommended that preoperative red cell transfusion was not necessary for laparoscopic surgery.[5-7] However; the subjects of our report differ in two very important respects. Firstly, the preoperative haemoglobin concentrations of our subjects were all below 4.0g/L in contrast to $\geq$ 6.0g/L in the reference studies. Secondly, our patients underwent open abdominal surgery which is potentially more associated with blood loss than laparoscopic surgery.

Given the status of the respiratory and heart rates, pulse oximetry and blood pressure; we have refrained from post-operative red cell transfusion, despite low haemoglobin levels. Instead, we reasoned that abrupt changes in their physiological status may not be suitable since the patients have spent several months with their very low levels of haemoglobin. Indeed, the physical growth of the subjects after surgery far exceeded pre-operative observations but also expectations of their otherwise well, non-HbSS peers, suggesting catch-up growth. The progressive increase in the haemoglobin levels and the catch-up pattern of physical growth observed in these cases provide new insights into the dynamic of erythropoiesis and organ function in children with HbSS. These observations highlight the positive changes that our CCCP can produce on HbSS children with severe presentation of the disease. We hope that the cases presented herein may help paediatricians convince anaesthetists and surgeons not to abandon some of the affected children presenting with massive chronic splenomegaly with hypersplenism and extreme low haemoglobin level.

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