Perioperative management for orthopaedic patients with sickle cell anaemia

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ABSTRACT

Purpose. To compare outcomes of 2 types of perioperative optimisation for patients with sickle cell anaemia (SCA) undergoing various orthopaedic surgeries.

Methods. 12 female and 11 male patients aged 13 to 40 (mean, 18) years with SCA underwent 31 separate orthopaedic procedures for osteonecrosis of the femoral head. They were referred to a haematologist for 2 types of perioperative optimisation, based on the choice of the attending paediatrician. In the aggressive management group, patients received packed red blood cells preoperatively to increase the haemoglobin level to 9 to 11 g/dl and to lower the haemoglobin S level to <30%. Fresh frozen plasma was given when their Factor VII level was <30%. In the conservative management group, patients received packed red blood cells preoperatively to increase the haemoglobin level to a minimum of 10 g/dl. Fresh frozen plasma or packed red blood cells were given intra-operatively only when excessive bleeding occurred. The length of hospital stay, the number of perioperative complications, the number of transfusions, and episodes of alloimmunisation and/or vaso-occlusive crises in the two groups were compared.

Results. No patient in the aggressive management group received supplemental oxygen or had an estimated intra-operative blood loss of >400 ml. Three patients in the conservative management group received multiple intra- and post-operative transfusions and supplemental oxygen.

Conclusion. Both aggressive and conservative protocols may be safely used in SCA patients. The more aggressive protocol resulted in lower rates for postoperative complications, transfusions, and resorting to supplemental oxygen.

Key words: anemia, sickle cell; blood component transfusion; blood transfusion; orthopedics; preoperative care

INTRODUCTION

Sickle cell anaemia (SCA) is a recessive genetic
disorder, in which there is a substitution of valine for glutamic acid in haemoglobin that renders red blood cells liable to sickling.\textsuperscript{1,2} It affects one of every 400 African-American newborns and approximately 70,000 people in the United States.\textsuperscript{3} Its causes of morbidity and mortality include vaso-occlusion and chronic haemolysis with resulting endothelial damage and ischaemic-reperfusion injuries.\textsuperscript{4,5} Patients with SCA are more likely to undergo cholecystectomies,\textsuperscript{6–11} splenectomies,\textsuperscript{10,12–14} tonsillectomies and adenoidectomies,\textsuperscript{15–17} and total hip arthroplasties.\textsuperscript{18–21} These patients are also at risk of multiple perioperative complications such as vaso-occlusive crises, haemolytic anaemia, acute chest syndrome, and multi-organ failure.\textsuperscript{1,3,5,19,22,23} About 20% of children with SCA develop intra-operative complications;\textsuperscript{22} intra-operative bleeding is also very common.\textsuperscript{24}

50% of adolescents and young adults with SCA developed osteonecrosis of the femoral head and many eventually undergo total hip arthroplasty (THA).\textsuperscript{25–27} This procedure requires prolonged anaesthesia and may involve a blood loss of >1 litre.\textsuperscript{21,26} Patients with SCA often have coagulation abnormalities, with elevated prothrombin times.\textsuperscript{29} Preoperative management of such patients differs among haematologists\textsuperscript{30,31}; it entails preoperative hydration, blood transfusions to decrease the percentage of haemoglobin S to <30%.\textsuperscript{5,9,30,32} Blood transfusion improves oxygen-carrying capacity and decreases the relative concentration of haemoglobin S, thus decreasing sickling and the risk of vaso-occlusive episodes. There is no consensus regarding perioperative optimisation of patients with SCA, particularly as to whether fresh frozen plasma should be given preoperatively or after bleeding occurs.\textsuperscript{30} Different stages of osteonecrosis of the femoral head necessitate different orthopaedic procedures and result in variable intra- and post-operative bleeding.\textsuperscript{30,34}

We compared outcomes of 2 types of perioperative optimisation for patients with SCA undergoing various orthopaedic surgeries.

**MATERIALS AND METHODS**

Approval for data analysis was obtained from our Institutional Review Board. Between January 2000 and December 2005, 12 female and 11 male patients aged 13 to 40 (mean, 18) years with SCA underwent 31 separate orthopaedic procedures for osteonecrosis of the femoral head. They were referred to a haematologist for 2 types of perioperative optimisation based on the attending paediatrician’s choice. The distribution of procedures was similar between the 2 groups (Table).

In the aggressive management group, patients received packed red blood cells preoperatively (as necessary) to increase the haemoglobin level to 9 to 11 g/dl and lower the haemoglobin S level to <30%. They were screened for coagulation disorders. Whenever the PT or partial thromboplastin time (PTT) were prolonged, preoperative fresh frozen plasma was given if Factor VII levels were <30%.

In the conservative management group, patients received packed red blood cells preoperatively to increase the haemoglobin levels to a minimum of 10 g/dl. Fresh frozen plasma or packed red blood cells were given intra-operatively only when excessive bleeding was deemed to occur. The PT and PTT were obtained in the course of regular preoperative workup, but no factor VII study was

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Aggressive management group</th>
<th>Conservative management group</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of males:females</td>
<td>6:9</td>
<td>6:2</td>
</tr>
<tr>
<td>Mean (range) age (years)</td>
<td>18 (13–40)</td>
<td>18 (14–35)</td>
</tr>
<tr>
<td>No. of procedures</td>
<td>16</td>
<td>15</td>
</tr>
<tr>
<td>Bone grafting</td>
<td>5</td>
<td>4</td>
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<tr>
<td>Core decompression</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>Iliopsoas releases with hip distraction</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Total hip arthroplasty</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Metal-on-metal hip resurfacing</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Mean preoperative haemoglobin level (g/dl)</td>
<td>11.3</td>
<td>10.4</td>
</tr>
<tr>
<td>Mean preoperative haemoglobin S (%)</td>
<td>29</td>
<td>52</td>
</tr>
<tr>
<td>Mean no. of units of blood transfused</td>
<td>3.8</td>
<td>2</td>
</tr>
<tr>
<td>No. of patients with prolonged prothrombin time</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>No. of patients with prolonged partial thromboplastin time</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>No. of patients with Factor VII level of &lt;30%</td>
<td>1</td>
<td>-</td>
</tr>
<tr>
<td>Mean (range) length of hospital stay (days)</td>
<td>2.3 (0–5)</td>
<td>3 (0–7)</td>
</tr>
<tr>
<td>Mean (range) intra- and post-operative blood transfusion (units)</td>
<td>1.2 (0–2)</td>
<td>2.1 (0–3)</td>
</tr>
<tr>
<td>No. of patients receiving packed red blood cells</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>No. of patients receiving supplemental oxygen</td>
<td>0</td>
<td>3</td>
</tr>
</tbody>
</table>
In both groups, criteria for intra- and post-operative transfusion were haemoglobin levels of <7 g/dl (haematocrit level of <21%), any episode of hypotension, refractory acidosis, or postoperative painful crisis. In addition to regular orthopaedic follow-up, specific examination for any adverse bleeding events were scheduled at month 1. The length of hospital stay, the number of perioperative complications, the number of transfusion, and episodes of alloimmunisation and/or vaso-occlusive crisis in both groups were compared.

Parametric analysis was performed with descriptive statistics. Comparison between groups was made using the 2-sample Student’s t-test. A p value of <0.05 was considered statistically significant. Owing to small sample size, comparisons were made in a descriptive manner.

RESULTS

Preoperatively, in the respective aggressive and conservative management groups, mean haemoglobin levels were 11.3 and 10.4 g/dl, mean haemoglobin S proportions were 29% and 52%, and mean units of blood transfused were 3.8 (5 patients had exchange and 2 had repeated transfusions) and 2 (6 patients had one unit each). Corresponding patient numbers with prolonged PT were 5 and 2, and one in each group had prolonged PTT. One patient who received exchange transfusion also had Factor VII levels of <30%. Intra- and post-operatively, the respective mean units of blood transfused were 1.2 (range, 0–2) and 2.1 (range, 0–3) [p=0.06, Student’s t-test]. One and 4 patients in the respective groups received 2 units of packed red blood cells. One THA patient in the conservative management group received 3 units of packed red blood cells (to counter estimated intra-operative blood loss of 1000 ml), 2 days of supplemental oxygen for respiratory compromise, and 7 days of hospitalisation. The outcome of this patient was good at the 2-year follow-up. Two additional patients in the same group received supplemental oxygen. No patient in the aggressive management group received supplemental oxygen or had an estimated intra-operative blood loss of >400 ml. The mean length of hospital stay (2.3 vs 3 days) in the 2 groups was not significantly different when adjusted for the stage of osteonecrosis and type of surgery. No patient had complications related to surgical failure or disease progression.

DISCUSSION

Blood loss management in patients with SCA is controversial. An aggressive preoperative transfusion protocol resulted in minimal peri- and post-operative adverse events and complications in 92 SCA patients aged one to 22 (mean, 10) years who underwent 130 surgeries. Of 32 SCA patients undergoing THA, 12 received transfusions (4 pre- and 8 intra-operatively) and 20 did not. All 4 patients receiving preoperative transfusions developed postoperative anaemia attributed to haemolysis. Three patients who did not receive transfusions developed acute chest syndrome, but none who were transfused did so. Nonetheless, some considered conservative transfusion regimens to be as effective as aggressive regimens in preventing perioperative complications in SCA patients, while resulting in only half as many transfusion-associated complications. 551 patients undergoing 604 elective surgeries were assigned to either an aggressive or conservative transfusion regimens, targeted to decrease the haemoglobin S level to <30% and increase the haemoglobin level to 10 g/dl, respectively. The respective serious complication rates were similar (31% vs. 35%), but transfusion-related complication rates differed significantly (14% vs. 7%). When orthopaedic procedures were matched, the difference became not significant. Administration of fresh frozen plasma or the perceived need for supplemental oxygen was not evaluated.

In our study, the sample size in various stages of osteonecrosis was small. SCA patients with osteonecrosis often undergo multiple surgical procedures during their lifetimes, and direct comparison is difficult. A randomised study is necessary to assess the advantages of different protocols.

CONCLUSION

Both aggressive and conservative transfusion protocols may be safely used in SCA patients. The more aggressive protocol resulted in lower rates for postoperative complications, transfusions and resorting to supplemental oxygen.

REFERENCES

1. Sathappan SS, Ginat D, Di Cesare PE. Multidisciplinary management of orthopedic patients with sickle cell disease.