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Perioperative Management in Children with Sickle Cell Disease Undergoing Laparoscopic Surgery

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ABSTRACT

Objective: The aim of this study was to evaluate our experience with laparoscopic surgery in children with sickle cell disease.

Methods: A retrospective chart review was performed to analyze the indication for surgery, perioperative management, surgical technique, complications, duration of hospitalization, and outcome. One pediatric surgeon performed all procedures.

Results: Thirteen children underwent laparoscopic surgery for the following indications: symptomatic cholelithiasis/cholecystitis in 9; recurrent splenic sequestration in 3; and hypersplenism/symptomatic cholelithiasis in 1. The 7 boys and 6 girls had a median age of 7.8 years. Patients undergoing splenectomy only were younger than those undergoing cholecystectomy (median age, 3.6 years versus 11.5 years, respectively). Four children underwent endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy because of common bile duct dilatation and stones. Twelve patients received packed red blood cell transfusions prior to surgery. The median operative time was 150 minutes, and the median hospitalization was 3 days. Four patients suffered postoperative complications (2 with acute chest syndrome, 1 with recurrent abdominal pain, and 1 with priapism). The patient with abdominal pain was found to have a retained stone in the common bile duct, which was retrieved via endoscopic retrograde cholangiopancreatography and sphincterotomy. All complications resolved with medical management.

Conclusions: Laparoscopic surgery is safe in children with sickle cell disease. Meticulous attention to perioperative management, transfusion guidelines, and pulmonary care may decrease the incidence of acute chest syndrome.

Key Words: Sickle cell anemia, Laparoscopic surgery, Child, Adolescence.

INTRODUCTION

Homozygous hemoglobin S (sickle cell) disease is a qualitative hemoglobinopathy whose clinical hallmarks are hemolytic anemia and vaso-occlusive/sickle cell crises.[1] Children so affected can suffer gallbladder disease in the form of cholecystitis and/or cholelithiasis and splenic sequestration. Cholelithiasis can cause severe bouts of abdominal pain and considerable morbidity. Splenic sequestration can rapidly progress to hypovolemic shock and death. Clearly, prompt medical attention is mandatory, especially for sequestration crises.

Cholecystectomy can cure gallbladder disease, and splenectomy will prevent subsequent splenic sequestration crises. However, children with sickle cell disease require meticulous perioperative care and long-term postoperative follow-up. Postsplenectomy bacterial infections can be quite severe with a possibly fatal outcome. Cholecystectomy should be performed as an elective procedure in all symptomatic patients with cholelithiasis, because emergency surgery during episodes of acute cholecystitis is associated with unacceptable morbidity. Splenectomy is best performed after the second sequestration crisis. With the advent of laparoscopic techniques, the need for postoperative analgesics and the length of hospitalization have been reduced.[2] The laparoscopic approach, moreover, may reduce the morbidity of surgery in children with sickle cell disease.

In the present report, we describe our single-institution experience with 13 children with sickle cell disease who underwent laparoscopic surgery for cholelithiasis and/or splenomegaly associated with recurrent splenic sequestration crises or hypersplenism.

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PATIENTS AND METHODS

We reviewed the charts of 370 children with sickle cell disease treated at Westchester Medical Center in Valhalla, New York, from 1995 to 2000. All are currently active patients in the pediatric hematology/oncology service. Thirteen children in this patient population were found to have undergone laparoscopic surgery. Nine of them underwent laparoscopic cholecystectomy because of symptomatic cholelithiasis; 3 underwent laparoscopic splenectomy because of recurrent splenic sequestration; and 1 underwent laparoscopic cholecystectomy/splenectomy because of symptomatic cholelithiasis and hypersplenism. Two of these patients have been previously described.[3] One pediatric surgeon (GS) performed all the laparoscopic surgeries. Patients undergoing splenectomy had preoperative abdominal ultrasound examinations to exclude cholelithiasis and had received at least 2 pneumococcal vaccinations. Four patients undergoing cholecystectomy had preoperative endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomies, and 2 underwent intraoperative cholangiograms to assist in identifying anatomic landmarks.

Operative Technique

The laparoscopic cholecystectomy was done by careful identification and isolation of the cystic duct and cystic artery. Two hemoclips were applied to the cystic duct proximal to the common bile duct, and 1 was applied distally. The junction of the cystic duct and common bile duct was always identified to prevent inadvertent damage to the common bile duct. Operative cholangiogram was performed only for specific indications. The gallbladder was removed from the gallbladder bed with monopolar or bipolar electrocautery. In the single case in which cholecystectomy was combined with splenectomy, the cholecystectomy was done first, with the patient in the supine position followed by the laparoscopic splenectomy (detailed below). An additional 5-mm port was added in the right upper quadrant for the cholecystectomy.

The splenectomy was performed with the patient in the right lateral decubitus position. This position allowed for the spleen to be suspended in the left upper quadrant by the splenic ligaments. At least 3 ports, as always required, were used for this procedure: a 5-mm port in the epigastrium, a 12-mm port in the left lower quadrant, and a 5-mm port in between. The 12-mm port was utilized to introduce the automatic endoscopic stapler (Endo GIA 30; US Surgical Corporation, Norwalk, CT) and subsequently to place the plastic specimen-retrieval bag (Endo Catch; US Surgical Corporation) to remove the spleen. A fourth port was needed in some cases to manipulate the spleen (it could be a mini 3-mm port or a 5-mm port), and it was placed in the left flank posteriorly to dissect behind the spleen. In most cases, the Harmonic scalpel was used (Ethicon, Johnson & Johnson Corporation, Somerville, NJ), but in a few cases, the bipolar cutting forceps (Everest Medical Corporation, Minneapolis, MN) were used. The use of surgical clips was kept to a minimum whenever possible, because they can interfere with the automatic stapler and cause bleeding.

The gastrosplenic ligament was divided at first. Initially, only the division of ligaments that was needed to expose the splenic hilum was performed. The remaining ligaments were preserved until the end of the operation to keep the spleen suspended in the left upper quadrant. The first branch of the blood supply from the gastroepiploic artery to the lower pole of the spleen was then divided. The splenic hilum was divided close to the spleen with the Endo GIA 30-2.0 vascular staples. Generally, 2 or 3 applications were necessary to completely divide the blood supply, including the short gastric vessels; in some cases, the remaining short gastric vessels could be divided with the Harmonic scalpel or the bipolar cautery. During application of the stapler, care was exercised to avoid the tail of the pancreas and the greater gastric curvature. After the blood supply had been divided, all the ligaments were transected with the bipolar forceps or Harmonic scalpel.

The spleen was placed inside the Endo Catch, and the opening of the bag was pulled out through the 12-mm port. The spleen was fragmented inside the bag with the finger or a ring blunt clamp and suction. This part of the procedure required careful attention to prevent rupture of the bag, which was watched from the inside. The spleen could be removed in small pieces. After the spleen was removed, care was taken to avoid contamination of the wounds or abdomen with splenic tissue to prevent splenosis.

At the beginning and at the end of the operation, a careful and methodical search was done to exclude accessory spleens. Long-acting local anesthesia (bupivacaine) was applied in all ports, and finally the ports were closed in the usual manner.

RESULTS

Thirteen children (7 boys and 6 girls) with sickle cell disease underwent laparoscopic surgery. The median age of the entire group was 7.8 years (range, 2.5 to 16.8 years); that of the splenectomy-only group was 3.6 years (range, 2.5 to 7.5 years), and that of the cholecystectomy group was 11.5 years (range, 7.8 to 16.8 years). The median operative time was 150 minutes (range, 90 to 310 minutes). The patient who underwent a combined cholecystectomy/splenectomy had an operative time of 300 minutes. In addition, 1 patient undergoing cholecystectomy required an intraoperative cholangiogram because of a difficult biliary anatomy and had an extended operative time of 310 minutes.

Nine patients received preoperative blood transfusions with a median of 10 cc/kg of packed red blood cells (range, 4 to 15 cc/kg). The median hemoglobin prior to transfusion was 8.4 g/dL (range, 6.1 to 9 g/dL), and the median hemoglobin posttransfusion was 9.8 g/dL (range, 9.2 to 13.2 g/dL). Three patients had received packed red blood cell transfusions 3 to 4 weeks prior to surgery: 1 for priapism refractory to intravenous hydration and analgesics, 1 for refractory pain during a vaso-occlusive crisis, and 1 for a second splenic sequestration crisis. One patient was not transfused. Her hemoglobin was 8.8 g/dL, and she did not suffer any perioperative complications.

The most common postoperative analgesic used was morphine sulfate. Eleven patients required a median of 24 hours of morphine sulfate (range, 24 hours to 6 days). One patient with postoperative acute chest syndrome and 1 with persistent abdominal pain required 4 and 6 days of intravenous morphine sulfate, respectively. One patient's postoperative pain was controlled with 24 hours of intravenous meperidine, and another patient required only acetaminophen with codeine. A second patient with postoperative acute chest syndrome required 5 days of acetaminophen and oxycodone after 24 hours of intravenous morphine sulfate.

The median postoperative hospitalization was 3 days (range, 2 to 12.4 days). The 4 patients who suffered postoperative complications (2 with acute chest syndrome, 1 with priapism, and 1 with persistent abdominal pain) had extended hospitalizations of 4, 6, 7, and 12 days. In addition to the pain management noted above, the patients with acute chest syndrome were treated with oxygen, intravenous hydration and antibiotics, and incentive spirometry. Diagnostic imaging in the patient with persistent abdominal pain showed a retained common bile duct stone and perihepatic fluid. An ERCP was performed that showed a dilated common bile duct and a stone in the distal common bile duct. The stone was extracted and a sphincterotomy performed.

Four patients underwent laparoscopic splenectomy (1 in combination with cholecystectomy) for recurrent splenic sequestration (n = 3) and 1 for hypersplenism. In 3 patients, the median splenic weight was 200 grams (range, 120 to 558 grams). One patient required a small incision in the left lower quadrant to assist in removing an 18 x 13 x 6-centimeter spleen weighing 558 grams.

The histopathology of the gallbladders showed chronic cholecystitis and cholelithiasis in each case. The histopathology of the spleens showed sinusoidal congestion with marked sickling erythrostasis. Two spleens had evidence of focal hemosiderosis, and in the 2 oldest patients (aged 7.5 and 7.8 years), evidence was present of atrophy of the splenic parenchyma.

No patient has had any long-term postoperative complications during a median follow-up time of 19 months (range, 1 month to 71 months). Two patients have required hospitalizations for recurrent vaso-occlusive crises, and 1 patient has required 2 admissions for postsplenectomy fever. In neither case did the patient with fever appear ill, and her hospital course was uneventful.

DISCUSSION

Our single-institution experience with laparoscopic surgery shows that it is a safe and effective approach to managing surgical problems in children with sickle cell disease. Three patients had complications related to sickle cell disease, and 1 patient had a retained stone in the common bile duct. Each complication was successfully managed, with no resultant deaths.

Cholelithiasis is a common clinical problem encountered in children with sickle cell disease,[1] and those with increased body mass index, reticulocyte counts, and levels of alkaline phosphatase are more likely to form stones.[4,5] Indeed, cholecystectomy is the most common surgical procedure performed on patients with sickle cell disease.[6] In pediatric patients with cholelithiasis, a careful search for choledocholithiasis should be undertaken,[7] and the presence of biliary sludge should prompt a referral to a pediatric surgeon.[8] However, in asymptomatic children with

cholelithiasis, observation seems prudent.[5]

With the advent of laparoscopic techniques, postoperative analgesia use and the duration of hospitalization have decreased. Eight studies in children with sickle cell disease and symptomatic cholelithiasis have shown that the technique is safe and effective.[9-16] The median hospitalization in these studies ranged from 1.6 to 2.3 days, and, when compared with open cholecystectomy,[10,14] was shorter for laparoscopic surgery. Most of the complications observed, especially acute chest syndrome, were related to sickle cell disease.

In our series, the median postoperative hospitalization was 3 days in the patients undergoing cholecystectomy. Interestingly, the 4 postoperative complications occurred in patients undergoing cholecystectomy (1 combined with splenectomy). The median operative time for these patients was longer than that for the patients undergoing cholecystectomy without postoperative complications (220 minutes versus 150 minutes, respectively). Perhaps the longer operative/anesthesia times of 300 and 310 minutes, in combination with a posttransfusion hemoglobin of 13.2 g/dL in 1 child, may have contributed to the development of the 2 cases of acute chest syndrome. It is noteworthy that, of 6 postoperative episodes of acute chest syndrome observed at another institution, 5 occurred after laparoscopic surgery and 1 after an open procedure.[17] Perhaps the benefits of laparoscopic surgery do not translate into a decreased incidence of acute chest syndrome; thus, meticulous perioperative and pulmonary care is mandatory.

Splenic sequestration is a principal cause of death in children with sickle cell disease.[18,19] Deaths result from hypovolemic shock. Recurrences are quite common among survivors of first attacks,[20] and most centers recommend splenectomy after the first recurrence.[21] Splenectomy does not increase the risk of death or bacteremic illnesses in children with sickle cell disease treated in this manner.[21] Indeed, splenectomy is safe and beneficial in children with sickling syndromes.[22]

In our series, all 3 patients undergoing splenectomy alone had an uneventful hospital course. Each child was vaccinated against pneumococcal disease and has been maintained on penicillin prophylaxis. One child with hypersplenism required a small incision in the left lower quadrant to facilitate removal of a large spleen. In 1 study, 17 children with sickling syndromes had splenectomy with massive splenomegaly (spleen weight of approximately 1000 grams).[23] Although the morbidity was slightly higher than that in patients with smaller spleens, the procedure was found to be generally safe. Newer techniques that are becoming available obviate the need for conversion to an open procedure, use of large and cumbersome intracorporeal bags, and the creation of additional incisions.

In our series, 9 patients received preoperative transfusions to attain a posttransfusion hemoglobin of 10 to 11 g/dL. One patient's posttransfusion hemoglobin was 13.2 g/dL, and this high level of hemoglobin, in addition to the prolonged operative/anesthesia time noted above, may have resulted in the case of acute chest syndrome that this patient developed. Current guidelines recommend that simple transfusion to increase the hemoglobin to 10 g/dL results in favorable postoperative outcomes.[24]

CONCLUSIONS

Laparoscopic surgery is the treatment of choice for children with sickle cell disease and symptomatic gallbladder disease, recurrent splenic sequestration, and hypersplenism. Sickle cell disease-related postoperative complications are the principal causes of morbidity. Meticulous attention to perioperative management, transfusion guidelines, and pulmonary status may further decrease the incidence of acute chest syndrome after laparoscopy.

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