Laparoscopic Splenectomy in Children with Sickle Cell Disease

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Background: Many children with Sickle Cell Disease (SCD) might experience severe sickle cell crises due to splenic complications. These include hypersplenism, acute splenic sequestration, splenic abscess and massive splenic infarction. Splenectomy is indicated to decrease the rate of recurrence of complications and the associated morbidity and mortality. The laparoscopic approach has proved to be associated with a better outcome. Many laparoscopic techniques were implemented for the removal of the spleen, especially when it is enlarged and fragile.

Objective: To evaluate laparoscopic assisted splenectomy technique and outcome in 51 children with SCD.

Design: A Retrospective Review.

Setting: Department of Pediatric Surgery, Salmaniya Medical Complex, Bahrain.

Method: Fifty-one children who had laparoscopic assisted splenectomy with a small inguinal incision from January 2002 to December 2014 were reviewed.

Result: Fifty-one children had laparoscopic assisted splenectomy for either hypersplenism 42 (82.4%) or acute splenic sequestration 9 (17.6 %); 32 (63%) males and 19 (37%) females. The age range was 6 to 14 years, a mean age of 9.8. Only one (1.9%) case required conversion to open procedure due to excessive bleeding. Only 7 (13.7%) were admitted in the ICU following the procedure. The mean length of hospital stay was four days; the measured decrease in the HBS was 38%, preoperative fever was seen in 16 (31%), and there was no mortality.

Conclusion: Laparoscopic splenectomy with a left inguinal incision is a safe and effective approach in children with SCD.

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Sickle Cell Disease (SCD) is a common genetic hematological disorder[1]. The spleen is one of the early organs to be affected by SCD. Early in life, splenomegaly occurs, and with repeated vaso-occlusion and infarctions, auto splenectomy would follow; although splenomegaly could persist in some patients[2]. Acute splenic sequestration, hypersplenism, splenic abscess and massive splenic infarction are serious SCD complications[2,3,4]. Treatment could be either conservative with repeat transfusions or surgical. Splenectomy is indicated due to the high risk of recurrence and increased morbidity and mortality of some of the above-mentioned complications[2].

The laparoscopic approach has been compared to open approach in multiple studies in children with SCD as well as other hematologic disorders, and has proven to be superior to the open approach[5-9]. It has the advantages of decreased pain, shorter hospital stay, fewer complications, faster return to normal activities and better cosmetic outcome[5-11]. Several techniques were practiced for laparoscopic splenectomy in children, some of which had been tried in SCD children because they require a rigorous management from their caregivers[10-17].

The aim of this study is to evaluate laparoscopic splenectomy technique and outcome in 51 children with SCD.

METHOD

Fifty-one patients who had laparoscopic splenectomy for SCD from January 2002 to December 2014 were reviewed. The age, sex, weight, indication for splenectomy, Sickle Hemoglobin percentage (HBS%) on admission and preoperatively, operative time, intraoperative complications, splenic weight, ICU admission, the length of hospital stay and conversion to open approach were documented. The inclusion criteria were patients with SCD below the age of 15, splenic complication, hypersplenism or acute sequestration. The following patients were excluded: adults or patients with other hematological disorders and patients needing concomitant cholecystectomy.

The diagnosis of SCD was based on Hemoglobin (HB) electrophoresis. Indications for splenectomy were either acute splenic sequestration or hypersplenism. Acute splenic sequestration crisis was defined as acute enlargement of the

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spleen or preexisting enlarged spleen with an evidence of red cell trapping within the spleen (acute anemia). Hypersplenism was defined as an increased splenic activity evident by persistent anemia, thrombocytopenia and/or neutropenia.

Adequate hydration was administered intravenously preoperatively. All patients received exchange transfusion to elevate the HB level to >11 g/dl and decrease the HBS% to < 50. All patients received antibiotics intravenously one day preoperatively until their discharge. Antibiotic choices were ceftriaxone or cefotaxime. All patients received pneumococcal, Haemophilus influenza and meningococcal vaccination according to the hospital management protocol for elective splenectomy. Abdominal ultrasound was performed preoperatively to screen for gallstones.

The patient was kept in the supine position. Foley catheter and nasogastric tube (NGT) were inserted. The surgeon was placed on the right of the patient. Four trocars were placed. Using the Hasson (open) technique, one 5 mm umbilical trocar was inserted for inflation with carbon dioxide. The other 5 mm ports were inserted: one in the left iliac fossa, one on the left anterior axillary line and one in the epigastric area just inferior to the xiphoid process. The stomach was moved away to expose the splenic pedicle and divide the splenocolic ligament to expose the splenic vessels. The splenic artery was identified and occluded with double surgical clips, followed by the splenic vein. The harmonic scalpel was used to divide the short gastric vessels and freely mobilize the spleen from the ligaments. An inguinal incision was made in the left side about half the width of the spleen size. The spleen was then introduced to the opening and was digitally fragmented to allow its extraction through the incision. Local peritoneal washing was performed. Trocars sites were closed with non-absorbable sutures (nylon sutures).

Patients known to have a history of acute chest syndrome were admitted to the ICU for close observation.

The patients were followed up for two weeks after discharge, and then three months and six months for a minimum of one year.

A student 'T' test was used to assess the significance of the difference between preoperative and postoperative HBS. Pearson correlation was used to assess the relationship between the splenic weight and the difference in HBS preoperatively.

RESULTS

The personal characteristics of fifty-one patients who had laparoscopic splenectomy are summarized in table 1. The mean age was 9.8 years (range 6-14), 32 (63%) males and 19 (37%) females. Forty-two (82.4%) were due to hypersplenism and 9 (17.6 %) splenic sequestration.

Table 1: Patients' Characteristics

Age	Mean ± SD	9.8 ±1.9
	Range	6-14
Weight (kg)	Mean ± SD	32.4 ± 6.9
	Range	21-50
Gender	Males	63%
	Females	37%

Table 2 shows the details of the measured outcome. The operative time varied from 75-190 minutes, a mean of 132 minutes. The reason for some prolonged procedures was the presence of extensive adhesions around the spleen. The spleen weight ranged from 50-470 grams, a mean of 131.7 grams. There is a weak significant positive correlation (r=0.29) between the weight of the spleen and the difference in HBS% preoperatively. There were no intraoperative complications except for one case of bleeding from the splenic artery, which required conversion to open approach and transfusion of one unit of blood. Seven (13.7%) cases were admitted to the ICU after the procedure for one day for close observation because they had history of acute chest syndrome. Specimens from all procedures were sent to the histopathology laboratory and reported sickling features. The mean hospitalization stay was four days, ranging from 2-7. The reason for prolonged hospitalization in 16 (31%) patients was pneumonia; all were treated empirically with proper antibiotics.

Table 2 : Measured Outcomes

Operative Time (minutes)	Mean ± SD	131.7 ± 30.6
	Range	75-190
Splenic Weight (grams)	Mean ± SD	218.8 ± 133.7
	Range	50-470
Length of Hospital Stay (days)	Mean	4
	Range	2-7
Need for ICU Admission	Percentage of Patients	13.7%
HBS% on Admission	Mean ± SD	63.9 ± 5.2
	Range	50-70
HBS% Preoperative	Mean ± SD	39.6 ± 5.1
	Range	30-55
Mean Decrease in HBS Preoperative	Percentage	38%
Intraoperative Complications-Bleeding	1 case only - percentage	1.96%
Conversion to Open		

The efficacy of splenectomy in children with SCD is evaluated by measuring the HBS percentage preoperative and postoperative. There was a decrease in HBS% in all patients, a mean decrease of 38% (P-value < 0.01). Nevertheless, there was no statistically significant difference between both genders in HBS% decrease on admission and preoperatively (P-value 0.9).

Other than pneumonia, there was no reported postoperative complication or mortality. No patient required postoperative blood transfusion or was readmitted after discharge. The follow-up revealed improved hematological and symptomatic profile.

DISCUSSION

Laparoscopic splenectomy in children was first described in 1993 by Tulmen et al[18]. Since then, it became increasingly used as a safe and effective procedure, and now is considered superior over open surgery[5-9]. Multiple advantages have been

linked to the laparoscopic approach; these include decreased pain, shorter hospital stay, better cosmetic outcome and faster return to normal activities[5-11]. The laparoscopic approach also has the advantage of available image magnification to allow clear visualization and full exploration of the abdominal cavity[19]. The disadvantages of laparoscopic approach are longer operating time and higher costs[14,20].

Several techniques were implemented for the laparoscopic removal of the spleen in children[10-17]. They vary in the patient's position, surgeon's approach, method of vessels devascularization and method of splenic extraction. The positions that were described for laparoscopic splenectomy include the supine position, the semi-lateral and the lateral. The approaches were anterior and lateral[15,21-22].

Different devices were tried for vessel devascularization; these include clips, sutures, ties, staplers, ultrasonic coagulation, monopolar and bipolar coagulation, Ethicon Endo-Surgery and ligature diathermy[15-16]. Splenic extraction could be performed using either an endo-bag, an electrochemical morcellator with the endo-bag or by doing a Pfannenstiel or an inguinal incision[12,15,19,22]. Although the endo-bag preserves the advantage of laparoscopic procedure as being minimally invasive, it could be difficult to manipulate and position the spleen correctly during extraction[22,23]. The electrochemical morcellator technique was described 17 years ago as a technique that allows the removal of a massively enlarged spleen via laparoscopy[12].

Pfannenstiel or inguinal incision were also described for splenic extraction although it may sacrifice the mini-invasiveness advantage of laparoscopy[23].

Hand assistance during laparoscopy as a method to decrease pulmonary complications was also described by Lee et al especially for enlarged spleens[24]. Nevertheless, this method was performed on adults, none of which had SCD.

Another technique described in the literature for splenectomy is the SIPES (single-incision pediatric endo-surgery)[25]. Three laparoscopic fascial incisions are performed, one of which is upsized for the endoscopic stapler and retrieval bag.

One alternative to both open and laparoscopic procedures, which preserves the mini invasiveness of laparoscopy is described by Esposito et al in 2008 as a mini-laparotomy incision in the subcostal region[24]. However, this approach does not have the cosmetic advantage of laparoscopy or lower groin incision, especially if the spleen is enlarged. In SCD children, it becomes more challenging when it comes to enlarged and fragile spleens.

Compared to other studies, our mean operative time is similar; however, our length of hospital stay is significantly higher[14,26]. This is attributed to the fact that SCD patients are at higher risk of complications than other patients[14].

Reported complications for both open and laparoscopic splenectomy include intraoperative and postoperative bleeding, incision infection, pancreatic leak, rupture of the diaphragm, subphrenic hematoma, rupture of the colon, portal or splenic vein thrombosis, splenic hematoma, postoperative fever, acute chest syndrome and pulmonary complications, particularly in SCD children[2,6,7,9,27,28]. In our study, only one case of intraoperative bleeding was converted to laparotomy (a conversion rate of 2%), and postoperative pneumonia in 31% of patients.

CONCLUSION

There is no standard procedure for laparoscopic splenectomy in children or particularly in children with SCD. Laparoscopic approach with a mini-inguinal incision is considered safe and effective for the removal of spleen in SCD children. Prospective blinded randomized controlled trial is recommended to compare the effectiveness of each laparoscopic approach for children with SCD.

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