

Laparoscopic Splenectomy in the Treatment of Childhood Hematologic Disorders

Esplenectomia Laparoscópica no Tratamento de Doenças Hematológicas da Infância

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ABSTRACT

Introduction: Spherocytosis, idiopathic thrombocytopenic purpura, and sickle cell disease are the most common indications for performing splenectomy in children. The aim of this study is to present our experience with laparoscopic splenectomy. **Material and methods:** A retrospective study was carried out analyzing the medical records of 86 patients. The following data were selected for analysis: age, sex, indications for splenectomy, presence of associated gallstones, presence of accessory spleens, surgical time, complications and hospital stay. **Results:** Age ranged from 6 months to 18 years with a mean age of 6.2. The weight ranged from 8 to 60 kg with a mean 23 kg. Splenectomy was performed by the following indications: sickle cell disease (45 cases), idiopathic thrombocytopenic purpura (23), Spherocytosis (11), thalassemia (3) autoimmune hemolytic anemia (1), myelodysplasia (1), Hodgkin's disease (1) and myelomonocytic leukemia (1). Cholelithiasis was diagnosed in six patients. Accessory spleens were found in 10 patients (12%). Bleeding was the more frequent intraoperative complication and in 5 patients (6%) caused the conversion to open surgery. Operative time ranged from 70 to 320 minutes with a mean of 160. Among early postoperative complications, one patient had pneumothorax and 3 had intra-abdominal fluid collection which resolved. Late complications included one patient with an umbilical incisional hernia and one patient with portal vein thrombosis. Hospital length-of-stay ranged from 2 to 21 days with an average of 3.2 and a median of 2 days. **Conclusion:** Laparoscopic splenectomy is a safe and effective alternative to open splenectomy.

Key words: Laparoscopy, splenectomy, portal vein thrombosis, children. Sickle cell.

Bras. J. Video-Sur, 2010, v. 3, n. 4: 195-199

Accepted after revision: August, 2010.

INTRODUCTION

Hematological disorders constitute the main indication for splenectomy in childhood. Spherocytosis, idiopathic thrombocytopenic purpura (ITP) and sickle cell anemia (SCA) make up the majority of cases. Splenectomy was performed exclusively via an open approach until 1991, when DELAITRE and MAIGNIEN performed the first laparoscopic splenectomy in an adult patient.¹ TULMAN and colleagues in 1993 performed the first laparoscopic splenectomy in children.² Since then, this technique has been gaining popularity due to accumulated experience and technological advances. The aim of this study is to present a single

service's experience with laparoscopic splenectomy in children.

METHODS

We conducted a retrospective study, analyzing the medical records of 86 children and adolescents who underwent elective laparoscopic splenectomy performed in Pediatric Surgery Service of the Darcy Vargas Children's Hospital in the period from July 2002 to March 2011.

The preoperative diagnosis and indications for splenectomy were established by the department of hematology and oncology, where patients were prepared for surgery and followed postoperatively.

Patients with a hemoglobin below 10 g/dl received packed red blood cells until the hemoglobin was between 10 and 12 g/dl. All patients received pneumococcal and meningococcal vaccines. The evaluation of patients with hemoglobinopathies included ultrasonography to assess the presence of associated cholelithiasis.

The laparoscopic technique involves inserting a 10 mm trocar in the umbilicus by the open technique. After creating the pneumoperitoneum, a 10 mm trocar is introduced in the left iliac fossa and two 5 mm trocars are placed, one subxiphoid and the other in the left anterior axillary line, below the rib cage. When cholecystectomy is also performed one more 5-mm trocar is employed in the right flank. The procedure begins with the opening of the gastrocolic ligament and extends along the entire gastric curvature and involves the short vessels. This maneuver exposes the pancreatic tail and splenic artery, which is ligated. Next the splenorenal and splenocolic ligaments are sectioned. Finally, the hilar vessels are sectioned after ligation with clips. Once completely freed, the spleen is placed into a plastic bag, crumbled into fragments, then eased out of the abdomen through a narrow incision.

The following variables were selected for analysis: age, sex, indication for splenectomy, presence of associated gallstones, presence of accessory spleens, surgical time, complications, and length of hospitalization.

RESULTS

Eighty six patients — 36 females and 50 males — underwent laparoscopic splenectomy. Age ranged from 6 months to 18 years with a mean of 6.2 and a median of 5.5 years. Weight ranged from 8 to 60 kg with a mean of 23 kg and median of 21 kg. Splenectomy was performed for the following indications: sickle cell anemia (45), idiopathic thrombocytopenic purpura (23), spherocytosis (11), thalassemia (3), autoimmune hemolytic anemia (1), myelodysplasia (1), Hodgkin's disease (1) and myelomonocytic leukemia (1). Cholelithiasis was diagnosed in six patients, who underwent concomitant cholecystectomy.

Accessory spleens were found in 10 patients (11%) and removed during the procedure. Bleeding was the most frequent intraoperative complication, and in 5 patients (6%) prompted conversion to open

surgery. Another case was complicated by opening of the diaphragm. The operative time ranged from 70 to 320 minutes with a mean of 160 and a median of 150 minutes.

Among the early postoperative complications, one patient had a pneumothorax that required chest tube drainage and three patients had intra-abdominal fluid collections that were treated conservatively.

The late postoperative complications included one patient who was noted to have an incisional umbilical hernia, one patient who developed portal vein thrombosis, and one patient with ITP who initially evolved with a partial response to the splenectomy, and was found on ultrasound to have an accessory spleen that had not been diagnosed intraoperatively. Later this patient was found to have a platelet count of around 160,000/uL; free of symptoms the patient was not re-operated. Inpatient length-of-stay ranged from 2 to 21 days, with a mean of 3.2 and a median of 2 days. Follow-up ranged from one month to 8 years and 9 months.

DISCUSSION

For nearly a century splenectomy has been used in the treatment of hematologic disorders. Classically performed by open surgery, starting in 1991 they began to be performed laparoscopically, initially in adults¹ and since 1993 in children.²

Laparoscopic splenectomy is a procedure of high complexity, whose technical difficulty is directly related to the presence of adhesions to other organs and the relative size of the spleen.

Adhesions occur involving the abdominal wall, omentum, stomach, colon and retroperitoneum, and are more frequent in sickle cell anemia with various crises and splenic sequestration and in cases of splenic abscess. When present, their lysis constitutes the first step of the surgery; despite the additional difficulty they do not constitute a contraindication to the procedure.

Splenomegaly is more common in spherocytosis and in lymphomas. The definition of the degree of splenomegaly in a quantitative manner through measurements is very inexact because of the wide age range in which the procedure is performed. Murawski and cols consider very marked splenomegaly, one in which the spleen reaches the height of the iliac crest.³ Splenomegaly until recently was considered a contraindication to the performance

of laparoscopy. Today this rule only applies to very pronounced splenomegaly that does not allow the mobilization of the organ and the visualization and exploration of the abdominal cavity in a safe manner.³ Less marked splenomegaly may be addressed by laparoscopy; a maneuver that usually improves the technical conditions is the prior ligation of the splenic artery, which results in “autotransfusion” and a decrease in the size of the spleen. In ITP, on the contrary, the spleen usually is normal in size. However, in the presence of corticosteroid treatment, there may be visceral fat accumulation which complicates the dissection of the splenic hilum.

In our series the main indication for splenectomy was for SCD with splenic sequestration crises, unlike the literature in which spherocytosis and ITP are the leading indications.⁴⁻⁶

The main intraoperative complication was bleeding, which in five cases led to conversion to open surgery.

Of the five patients, three had sickle cell disease and two had spherocytosis. All presented with splenomegaly; those with SCD also had extensive adhesions. The rate of conversion to open surgery was 6% (n = 5), which compares with a range of 2 to 18% in the global literature.⁴⁻¹⁰

The identification and removal of accessory spleens is another important issue, since missing them may allow the condition for which the splenectomy may persist or there may later be a recurrence of symptoms. The incidence of accessory spleens was 12% in our sample, whereas in other series this rate varies from 4 to 28%.^{6,7,10-14} The vast majority are located in the region of the tail of the pancreas, omentum and splenic hilum. The laparoscopic approach seems to imply an increased difficulty in locating them and they should be removed as soon as they are identified.

Surgical time is related to the learning curve, material available, anatomical conditions such as the size of the spleen and the presence or absence of

adhesions. In our series the average was 160 minutes. On the other hand, the median duration of the postoperative hospital stay was 48 hours.

A review of the literature reveals that these two variables – surgical time and duration of the hospitalization – represent the most conflicting results when analyzing the open approach versus the laparoscopic technique. However, these authors^{6,8,10,11} are unanimous in asserting that the longer duration of laparoscopic surgery is offset by a lower postoperative morbidity, a shorter hospital stay, a quicker return to normal activities, an earlier return to other treatments were postponed, as well as better aesthetic results.

The main postoperative complication occurred in one patient with spherocytosis who developed portal vein thrombosis (PVT), which was diagnosed about one year after the surgery when the patient experienced upper gastrointestinal hemorrhage due to bleeding esophageal varices. The estimated incidence of symptomatic PVT is between 0.07% and 2%, but the incidence found on routinely performed imaging studies can be between 7% and 10%. It is more frequent in adults. It is characterized by vague symptoms such as fever, vomiting, abdominal pain. DVT occurs in adults on average on the sixth day after splenectomy, but the period of time can vary up to 18 months. The occurrence of these symptoms after surgery involves the performance of Doppler ultrasound of the portal venous system. DVT occurs most frequently in patients suffering from myeloproliferative disorders, hemolytic anemia with splenomegaly and thrombocytosis large. Thrombosis occurs as frequently in open splenectomies as with laparoscopic splenectomies.¹⁵⁻¹⁸

CONCLUSION

Laparoscopic splenectomy is a safe and effective alternative to open surgery in the treatment of hematologic malignancies of childhood.

RESUMO

Introdução: Esferocitose, purpura trombocitopênica idiopática e anemia falciforme são as indicações mais comuns para a realização de esplenectomia em crianças. O objetivo desse estudo é apresentar a experiência com o uso da esplenectomia laparoscópica. **Métodos:** Foi realizado um estudo retrospectivo analisando os prontuários de 86 pacientes. Os seguintes dados foram coletados para análise: idade, sexo, indicação da esplenectomia, presença de colelitíase associada, presença de baços acessórios, duração da cirurgia, complicações e permanência hospitalar. **Resultados:** A idade variou de 6 meses a 18 anos com uma média de 6,2. O peso variou de 8 a 60 kg com uma média de 23 kg. A esplenectomia foi realizada pelas seguintes indicações: anemia falciforme (45); purpura trombocitopênica idiopática (23); esferocitose (11); talassemia (3); anemia hemolítica auto-imune (1); mielodisplasia (1), Doença de Hodgkin (1) e leucemia mielomonocítica (1). Colelitíase foi diagnosticada em 6 pacientes. Baços acessórios foram encontrados em 10 pacientes. O sangramento foi a complicação intra-operatória mais freqüente, sendo que em 5 pacientes (6%) determinou a conversão para cirurgia aberta. O tempo operatório variou de 70 a 320 minutos com média de 160. Entre as complicações pós-operatórias precoces, um paciente apresentou pneumotórax e 3 pacientes apresentaram coleção intra-abdominal. Entre as complicações pós-operatórias tardias, foi registrado um paciente com hérnia umbilical incisional e um paciente com trombose de veia porta. O tempo de internação variou de 2 a 21 dias com média de 3,2 e mediana de 2 dias. **Conclusão:** A esplenectomia laparoscópica é uma alternativa segura e eficaz no tratamento das doenças hematológicas da infância.

Descritores: Laparoscopia, esplenectomia, trombose veia porta, anemia falciforme, criança.

REFERENCES

1. Delaitre B, Maignien B. Splenectomy by the laparoscopic approach: report of a case. *Presse Med.* 1991; 20: 2263.
2. Tulman S, Holcomb GW, Karamanoukian HL, Reinhout J. Pediatric laparoscopic splenectomy. *J Pediatr Surg.* 1993; 28: 689-92.
3. Murawska M, Patkowskib D, Korlackic W, Czauderna P, Srokaa M, Makarewiczd W, Czernikb J, Dzielickic J. Laparoscopic splenectomy in children—a multicenter experience. *J Pediatr Surg.* 2008; 43: 951-954.
4. Rescorla F J, West KW, Engum AS, Grosfeld JL. Laparoscopic splenic procedures in children: experience in 231 children. *Ann Surg.* 2007; 246(4): 683-8.
5. Esposito C, Schaarschmidt K, Settimi A, Montupet Ph. Experience With Laparoscopic Splenectomy. *J Pediatr Surg.* 2001; 36(2): 309-11.
6. Murawski M, Patkowskib D, Korlackic W, Czauderna P, Srokaa M, Makarewiczd W et al. Laparoscopic splenectomy in children –a multicenter experience. *J Pediatr Surg.* 2008; 43: 951–4.
7. Chowbey PK, Goel A, Panse R, ANIL Sharma A, Khullar R, Soni V et al. Laparoscopic splenectomy for hematologic disorders: Experience with the first fifty patients. *J Laparoendoscopic Adv Surg Tech.* 2005; 15(1): 28-32.
8. Owera A, Hamade AM, MD, Hani OIB, Ammori BJ. Laparoscopic versus open splenectomy for massive splenomegaly: A comparative study. *J Laparoendoscopic Adv Surg Tech.* 2006; 16(3): 241-6.
9. Palanivelu C, Jani K, Malladi V, Shetty R, Senthilkumar R, Maheshkumar G. Early ligation of the splenic artery in the leaning spleen approach to laparoscopic splenectomy. *J Laparoendoscopic Adv Surg Tech.* 2006; 16(4): 339-44.
10. Patkowski D, Chrzan R, Wróbel G, Sokól A, Dobaczewski G, Apoznan'ski W et al. Laparoscopic splenectomy in children: Experience in a single institution. *J Laparoendoscopic Adv Surg Tech.* 2007; 17(2): 230-4.
11. Minkes RK, Lagzdins M, Langer JC. Laparoscopic versus open splenectomy in children. *J Pediatr Surg.* 2000; 35(5): 699-701.
12. Danielson PD, Shaul DB, Phillips JD, Stein JE, Andreson DA. The technical advances in pediatric laparoscopy have had a beneficial impact on splenectomy. *J Pediatr Surg.* 2000; 35(11): 1578-81.
13. Katkhouda N, Manhas S, Umbach TW, Kaiser AM. Laparoscopic splenectomy. *J Laparoendoscopic Adv Surg Tech.* 2001; 11(6): 383-90.
14. Kucuk C, Souzer E, Ok E, Altuntas F, Yilmaz Z. Laparoscopic versus open splenectomy in the management of benign and malign hematologic diseases: a ten year single Center experience. *J Laparoendoscopic Adv Surg Tech.* 2005; 15(2): 135-9.
15. Brink JS, Brown AK, Palmer BA, Moir C, Rodeberg DR. Portal vein thrombosis after laparoscopic assisted splenectomy and cholecystectomy. *J Pediatr Surg.* 2003; 38(4): 644-7
16. Ođuzkurta P, Tercanb F, Ýncea E, Ezera SS, Akgün Hiçsönmeza. Percutaneous treatment of portal vein thrombosis in a child who has undergone splenectomy. *J Pediatr Surg.* 2008; 43: E29-E32.
17. Soyer T, Ciftci AO, Tanyel FC, Senocak ME, Bu`yu`kpamukc N. Portal vein thrombosis after splenectomy in pediatric hematologic disease: risk factors, clinical features and outcome. *J Pediatr Surg.* 2006; 41: 1899-902.

18. Rossi E, Michelini ME, Pignatti CB, Zanotti F, Franchella A. A case of portal vein thrombosis after laparoscopic-assisted splenectomy and cholecystectomy in a child. *J Pediatr Surg.* 2007; 42: 1449-51.

ABREVIATIONS

ITP - Idiopathic Thrombocytopenic Purpura

FA - Falciform Anemia

PVT - Portal Vein Thrombosis

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