Surg Endosc (2006) 20: 748–752
DOI: 10.1007/s00464-005-0429-6

SURGICAL ENDOSCOPY
and Other Interventional Techniques

Laparoscopic subtotal splenectomy in hereditary spherocytosis

To preserve the upper or the lower pole of the spleen?

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Received: 16 June 2005/Accepted: 26 November 2005/Online publication: 16 March 2006

Abstract
Background: Clinical manifestations of hereditary spherocytosis can be controlled by splenectomy. The use of this procedure has been restricted due to concerns regarding exposure of patients to a lifelong risk of overwhelming infections. Subtotal splenectomy, which removes 85–90% of the enlarged spleen, is a logical alternative. In the first cases performed by laparoscopy we have chosen to preserve the upper pole. However, this technique showed some disadvantages, especially concerning the correct intraoperative evaluation of the splenic remnant volume. Therefore, we developed a new variant of the procedure by preserving the lower pole of the spleen.
Methods: Based on the authors' experience in laparoscopy (176 laparoscopic splenectomies), 10 laparoscopic subtotal splenectomies were performed in patients with hereditary microspherocytosis, preserving either the upper or the lower splenic pole.
Results: Patient age ranged between 5 and 35 years. The mean volume of the remnant spleen was 41.4 cm^3. There were no complications, and no transfusions were needed. Follow-up for 1–30 months was available.
Conclusion: Subtotal splenectomy appears to control hemolysis while maintaining splenic function. The laparoscopic approach is safe and effective and should be considered the procedure of choice in hereditary microspherocytosis. Laparoscopic subtotal splenectomy presents an advantage over open subtotal splenectomy, resulting in decreased blood loss, shorter hospital stay, no conversions, fewer operative and postoperative complications, and excellent remission rates. On the basis of our experience, the preservation of the lower pole of the spleen seems to be a first-line option for the optimal evaluation of the residual splenic mass.

Key words: Subtotal splenectomy — Partial splenectomy — Subtotal laparoscopic splenectomy — Hereditary spherocytosis — Laparoscopic splenectomy

Hereditary spherocytosis (HS) is the most common inherited hemolytic anemia in which a primary deficiency in an erythrocyte membrane skeleton protein leads to surface area loss. Erythrocytes with decreased surface area and impaired deformability are trapped in the splenic pulp and phagocytized [19]. The result is chronic hemolysis and an increased propensity for gallstone formation.

The clinical severity of HS varies from symptom-free carrier to severe hemolysis. Patients with HS should be graded by their severity of disease (baseline hemoglobin, reticulocyte count, and jaundice level of activity). This predicts the clinical course and the need for splenectomy [4].

Splenectomy is very effective in reducing hemolysis, leading to a significant prolongation of the red cell life span. The clinical manifestations and complications (anemia and gallstones) are much reduced in severe HS and abolished in milder cases, but at the price of an increased risk of life-threatening sepsis, usually caused by pneumococcal species. This risk is age related, being highest in the youngest children and within the first years after surgery.

Current guidelines recommend pre-splenectomy vaccination against pneumococcus, haemophilus, and meningococcus together with long-term (lifelong)

The paper is based on material presented at the 13th International Congress of the European Association for Endoscopic Surgery (EAES), Venice, 2005

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postoperative penicillin prophylaxis. These measures do not completely eliminate the risk because of the serotypes that are not represented in vaccines, penicillin-resistant pneumococcal strains, and issues relating to lifelong compliance. Recent studies have raised additional concerns regarding long-term complications after splenectomy, such as atherosclerotic events and pulmonary hypertension [2, 19].

Overwhelming postsplenectomy infections (OPSI) are primarily caused by *Streptococcus pneumoniae*, *Neisseriae meningitides*, or *Haemophilus influenzae*. Despite appropriate antibiotics and intensive therapeutic intervention, the overall mortality in published studies for established cases of OPSI varies from 50 to 70%. Of those patients who die, > 50% die within the first 48 h of hospital admission. In those patients who survive, other sequelae include gangrene leading to amputations, deafness associated with meningitis, or mastoid osteomyelitis and aortic insufficiency secondary to endocarditis [5].

Open subtotal splenectomy has been proven to be a therapeutic alternative in HS. In order to add the advantages of minimally invasive surgery, we began to perform this procedure by laparoscopy. Based on the authors' experience in laparoscopic surgery (176 laparoscopic splenectomies) and after performing five open subtotal splenectomies with good hematological outcomes, we performed laparoscopic subtotal splenectomy in 10 patients. In open surgery, subtotal splenectomy was performed preserving either the upper [11] or the lower pole of the spleen [2, 16, 18]. In the first cases [22] performed by laparoscopy, we chose to preserve the upper pole. However, this technique showed some disadvantages, especially concerning the correct intraoperative evaluation of the splenic remnant volume. Therefore, we developed a new variant of the procedure by preserving the lower pole of the spleen.

Patients and methods

Patients

Between June 2002 and January 2005, 10 consecutive patients underwent laparoscopic subtotal splenectomy. The patients were six children (four girls and two boys; age range, 5–12 years; mean age, 7.5 years) and four young adults (age range, 19–35 years). The patients' characteristics are detailed in Table 1. The body mass index ranged between 17 and 31. The diagnosis was based on clinical features (variable degree of anemia, jaundice, and splenomegaly) and laboratory findings (spherocytes, raised red cell hemoglobin concentration, and an increase in reticulocytes). Spleen size and volume were determined using ultrasounds. Spleen volume ranged between 243 and 760 cm^3 (mean, 429 cm^3). When small accessory spleens were detected they were preserved. The indication for surgery was severe anemia with recurrent need for transfusions, and surgery was performed after clinical monitoring of a number of variables (the degree of anemia, the impact of chronic fatigue on life quality, and the presence of gallstones). After discussions with the parents or the patients and after explaining the potential benefits and risks, subtotal laparoscopic splenectomy was performed. The procedure was feasible in all patients, and at the time of the intervention none required final total splenectomy. The mean duration of follow-up for the 10 patients was 13.1 months (range, 1–30).

Surgical technique

The objective of the surgical procedure was to remove approximately 85% of the splenic tissue while preserving either the upper or the lower pole of the spleen. Based on the authors' experience and standard approach in laparoscopic splenectomy, the upper pole of the spleen was preserved in the first seven procedures according to the recommendations of Petroianu et al. [13]. In the last three cases, the lower pole of the spleen was preserved based on our experience in open surgery (five open subtotal splenectomies). Under general anesthesia, the patient was placed in the right lateral decubitus position. Four trocars were used, two 12 mm ports (one for the laparoscope and the other for the hemostatic instruments) and two 5 mm ports for retractors. Trocar position was similarly utilized with any laparoscopic splenectomy. A 30° laparoscope was inserted into the abdominal cavity and a limited diagnostic laparoscopy was performed in order to detect accessory spleens (an accessory spleen was observed in one case and was preserved).

Subtotal laparoscopic splenectomy preserving the upper pole

Initially, the splenocolic and phrenosplenic ligaments were partially incised using a harmonic scalpel, and the branches of the gastroepiploic vessels to the lower pole were divided (Fig. 1A). The splenic hilum was carefully dissected, preserving the pancreatic tail intact. The branches of the splenic artery and vein were divided using Ligasure Atlas Sealer/Divider (Tyco Healthcare, Boulder, CO, USA). The last one or two short gastric vessels were preserved by observing the size of the future splenic remnant, which provided a clear line of demarcation on the spleen surface. Standard monopolar electrocautery was used to transect the spleen, ensuring that a 5 mm rim of devascularized splenic tissue remained in situ. Monopolar electrocautery was used in favor of the Ligasure or the harmonic shears in order to visualize splenic remnant vascularization. After that, the Ligasure Atlas Sealer/Divider provided excellent hemostatic control. A TachoComb (Nycomed GmbH, Linz, Austria) was used to complete haemostasis. The splenic fragment was placed in an endobag for removal. The most lateral 12 mm port site was used to extract the morcellated tissue, or the specimen was extracted via a Pfannenstiel incision. Two drains were placed at the end of the procedure. The postoperative courses were uneventful, and all patients were discharged within 1 week after surgery.

Subtotal laparoscopic splenectomy preserving the lower pole

This technique is similar to that used in the open approach (Fig. 1B). After dividing the branches of the splenic hilum and the short gastric vessels using Ligasure Atlas Sealer/Divider, a small fragment of the lower pole was preserved. The vascularization was based on the ascending branch of the left gastroepiploic artery. The advantage of this technique was better visualization of the remnant size.

Results

The mean surgery time was 95 min. The mean operative bleeding was < 70 ml. No intraoperative or postoperative blood transfusions were required. In six cases, the specimen was extracted using an endobag after morcellation. In these cases, weight of the specimen was calculated using the following formula: intact weight (g) = morcellated weight (g) × 1.34 + 45 [23]. In four cases, the specimen was extracted via a Pfannenstiel incision. The mean weight of the resected specimen was 600 g.

Table 1. Demographic data of patients and values of hemoglobin and reticulocytes before and after surgery

Patient No.	Age at surgery (yr)	Transfusions before surgery	Spleen volumetry before/after surgery (cm^3)	Hemoglobin value (g/dl)		Reticulocyte value, post-splenectomy (%)	Postsurgical follow-up (mo)
				Pre-splenectomy	Post-splenectomy		
1	16	Yes	664/71	8.4	11	16.2	32
2	19	Yes	760/79	7.7	11	12.7	31
3	5	Yes	336/35	8.6	10.7	18.5	20
4	35	Yes	603/80	8	10.2	11.3	19
5	7	Yes	307/21	7.5	9.4	8.7	13
6	5	Yes	373/32	8	10.6	6.3	13
7	8	Yes	317/29	8.9	11.8	19.6	12
8	12	Yes	278/21	7.4	10.4	8.7	4
9	22	Yes	408/26	8.9	12.1	9.8	4
10	10	Yes	243/20	7.6	9.5	6.3	3

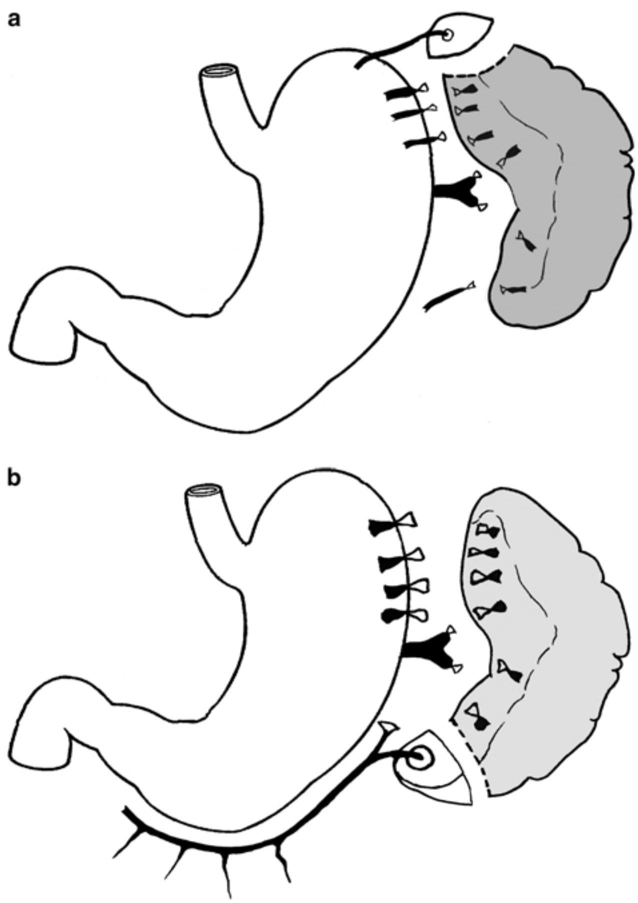

Fig. 1. (**A**) Surgical technique used for laparoscopic subtotal splenectomy: the remnant upper pole of the spleen with its vascular supply (a short gastric vessel). (**B**) Surgical technique used for laparoscopic subtotal splenectomy: the remnant lower pole of the spleen with its vascular supply (a pedicle arising from the left gastroepiploic vessels).

In all patients, the hemoglobin values were increased and the reticulocyte number was decreased following subtotal splenectomy (Table 1), as a direct consequence of an increase in the life span of red blood cells. During a follow-up period that ranged from 1 to 30 months, none of the patients required blood transfusions to maintain adequate hemoglobin levels. In all patients, a Doppler ultrasound performed after surgery showed the vascularization of the splenic remnant. Comparison of the spleen volumetry before and after surgery showed that the splenic remnant represented 15% of the initial volume of the spleen. Following Stoehr et al. [18], we assessed the phagocytic activity of the remnant spleen by scintigraphy with 99-technetium, which showed radionuclide uptake in the splenic area. Also, the percentage of pitted erythrocytes was < 2%. Howell–Jolly bodies appeared transiently after surgery in six of 10 patients. One patient required splenic remnant removal 11 months after the initial surgery due to persistent mild hemolytic anemia. In this case, due to the adhesions of the splenic remnant, conversion to open surgery was necessary.

Discussion

Indications for surgical treatment

Splenectomy should be performed in children with severe transfusion-dependent HS. The clinical manifestations of HS include anemia, chronic jaundice, and cholelithiasis. Total splenectomy that abrogates hemolysis is the most attractive treatment option, but this exposes the patient to a life-long risk of potentially lethal infections. Splenectomy leads to a significant prolongation of the red cell life span (although this is not necessarily extended to a normal life span). By eliminating the need for regular blood transfusions in severe cases, this surgical procedure also eliminates the associated risks of blood-borne viral infections and iron overload [20]. In addition, the impact on the patient's quality of life is significant, with a major improvement in the patient's physical and professional abilities after surgery.

Unfortunately, spleen removal leads to many inconveniences because the spleen represents approximately 25% of the lymphatic tissue and is the major repository of mononuclear phagocytic cells. As a result, it performs an efficient filtration function for the bloodstream [14]. OPSI is a well-defined clinical condition resulting from the inability of the splenectomized host to combat infections with encapsulated bacteria. Because of this lethal complication, a number of alternatives to splenectomy have been studied, including autotransplantation of the removed spleen and partial splenectomy [1, 6, 14]. Partial splenectomy is preferable to splenic autotransplantation because it is associated with higher antibody titers after immunization, better pneumococcal splenic uptake, and improved survival rates.

Subtotal splenectomy provides a potentially effective alternative to total splenectomy in patients with HS. It is likely that contact between antigens and lymphoid cells as well as maturation of the pre-B cells can proceed in the remnant spleen.

In the study by Bader-Meunier et al. [2], approximately 15% of the enlarged spleen was preserved in order to decrease the sequestration of red blood cells as much as possible while at the same time maintaining an adequate amount of remnant spleen to preserve its phagocytic function.

Key points in the surgical technique

Open subtotal splenectomy has been suggested as a feasible alternative for severe forms of HS [2, 16, 18]. The potential benefits of this method were first proposed by a group of French surgeons, who published between 1993 and 2001 a study based on 40 cases. Since then, the technique has been applied by others, and additional studies have shown that this method provides good results [16]. It has also recently been hypothesized that a more radical approach to open resection could permanently decrease recurrent hemolysis and at the same time conserve a remnant spleen of 10 cm^3 [18]. Seshadri et al. [17] performed a partial laparoscopic splenectomy for a 2 cm splenic cyst. In the current study, after five open subtotal splenectomies were performed with good hematological outcomes, the potential benefits of the laparoscopic approach were analyzed because laparoscopic splenectomy is currently considered the best approach in patients without splenomegaly [22].

The development of partial splenectomy is based on the knowledge of the splenic vascular pattern [13, 21]. This surgical approach is feasible because of the unique anatomical features of this organ, which is divided into a variable number of almost independent vascular territories. Splenic vessels, including branches of the main splenic pedicle, short gastric vessels, and left gastroepiploic vessels, are divided outside the spleen before entering the splenic hilum as terminal vessels. There are three types of anastomoses between the individual branches of the splenic artery: hilar, intraparenchymatous, and subcapsular [3, 7, 8]. The intraparenchymatous anastomoses do not maintain the viability of the externally devascularized regions of the spleen, a fact that makes it possible to obtain a safe hemostasis of the splenic remnant. Subtotal splenectomy can be performed after primary ligation of the main pedicle of the splenic artery and vein, preserving either a short gastric artery [10] or a branch of the left gastroepiploic artery [20].

In order to establish the best therapeutic option, it is essential to determine the amount of splenic parenchyma that needs to be removed to effectively reduce red blood cell destruction. In addition, it is important to evaluate the minimal amount of residual parenchyma necessary for maintaining adequate phagocytic function [2, 19, 20]. Previous studies have shown that this goal can be achieved by preserving approximately 15% of the enlarged spleen. This is the key point of the surgical procedure.

In the current study, the technique described by Tchernia et al. [2, 19, 20] was used during open surgery to preserve the ascending branch of the left gastroepiploic artery. This solution was initially difficult to apply in the laparoscopic approach because in this case the procedure is conducted after manipulation of the lower splenic pole. As a result, Petroianu's technique was initially used as a part of the laparoscopic approach. This technique has been used successfully in a large number of operations to preserve the splenogastric vessels, which are considered to be enough for the upper splenic pole [9, 11, 12, 15].

In the current study, the lower splenic pole was preserved in the last three patients with good results. In the first procedures, according to the recommendation of Bader-Meunier et al. [2], approximately 15% of the enlarged spleen was preserved, a target that was more difficult to achieve using the laparoscopic approach than with open surgery. In the first procedures the main difficulty was to approximate 15% of the enlarged spleen. The evaluation of the splenic remnant volume was quite difficult in the cases with preservation of the upper pole of the spleen. Therefore, in the last cases we chose to preserve the lower pole of the spleen. This technique allowed us a better mobilization of the splenic remnant and a more accurate evaluation of the volume. The length of the remnant ranged between 3.5 and 6 cm, depending on the initial size of the spleen, and was measured using a 10-cm flexible band. Based on the 10 cases in this study and recent publications [18], it appears that a smaller remnant spleen (a remnant volume between 5 and 10%) is sufficient to preserve the splenic immune and phagocytic functions. Laparoscopic subtotal splenectomy combines the advantages of minimally invasive surgery with the maintenance of phagocytic function of the splenic remnant.

Hematologic results

The hemoglobin values increased and the reticulocyte count decreased in the first several days after surgery. This was the result of increased bone marrow production, evidenced by 20% reticulocytes.

Studies published between 1993 and 2001 [2, 19, 20] based on 40 cases that were followed for 14 years after surgery showed that the mean increase in hemoglobin value after partial splenectomy was 3 g/dl, whereas the decrease in the number of reticulocytes averaged approximately 300×10^9/L [2]. This occurred as a direct result of an increase in the red blood cell life span, with an average increase of 6.5 $\pm$ 1 days—a result far from normal.

Long-term postsurgical follow-up results show that beneficial clinical effects are sustained over a long period. However, in one study, one patient experienced complications that led to a secondary total splenectomy [2]. A male patient who underwent surgery at the age of 5 years and maintained a hemolytic state after surgery required a secondary splenectomy (clinical symptoms included recurrence of jaundice, chronic fatigue, and mild anemia). Laparoscopic surgery was attempted, but it was necessary to convert to open surgery due to phrenosplenic adhesions. Other complications that also require secondary splenectomy include bleeding and torsion of vascular pedicles.

Several studies found that secondary splenectomy was required in 10% of patients at 5 years and in 33% at 10 years [2, 20]. The size of the remnant spleen increased in all cases. The growth was assessed by ultrasound and CT scan. Significant regrowth of the splenic remnant was noted during the first year after surgery, sometimes four and a half times its postoperative size [18], which did not correlate with the measured hemoglobin values or reticulocyte counts [2]. Although the current study is based on a small series of patients and the follow-up is short, our aim was to present a new approach in patients with HS. Of course, long-term results are needed in order to prove that the effects are sustained over a long period.

The outcome of patients with HS with total splenectomy was compared to that of patients with subtotal splenectomy [20] performed by open surgery. The results showed that subtotal splenectomy appears to be the first-line treatment option for patients with HS [2]. Regarding the phagocytic activity of the remnant, the percentage of pitted cells after partial splenectomy was similar to that seen in normal individuals, whereas after total splenectomy their numbers were markedly increased [20].

Conclusions

Subtotal splenectomy has been shown to provide a persistent decrease in hemolytic rate while preserving the integrity of splenic phagocytic function. Subtotal splenectomy should be considered preferable in two situations. First, in children younger than 5 years of age who need regular transfusions, subtotal splenectomy decreases the risk of both severe sepsis and transfusion-related viral infections. Second, subtotal splenectomy should be considered for cases in which mild or moderate anemia is present and the patient has chronic discomfort, significant splenic enlargement, or biliary lithiasis but does not require transfusions or requires them only sporadically. Laparoscopic subtotal splenectomy presents an advantage over open subtotal splenectomy, resulting in decreased blood loss, shorter hospital stay, no conversions, fewer operative and postoperative complications, and excellent remission rates. On the basis of our experience, the preservation of the lower pole of the spleen seems to be a first-line option for the optimal evaluation of the residual splenic mass. The laparoscopic approach is safe and effective, with encouraging short-term results, but a much longer follow-up (up to 20 years) is needed to be certain that the beneficial effects will be sustained.

Acknowledgment. We thank Leslie Cizmas, Ph.D., Texas A&M University.

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