

EFFICACY OF LAPAROSCOPIC SPLENECTOMY IN THALASSEMIA PATIENTS

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Abstract

Thalassemia is one of the most common monogenic disorders in India. Approximately 7500- 12,000 babies with β -thalassemia major are born every year in the country. [13] Because of the magnitude of the disease and health-care burden it poses, it has been covered as part of various National Programs by the Government of India. Children with features of hypersplenism and increased packed red blood cell (PRBC) transfusion requirement are referred to the pediatric surgical unit for splenectomy. Open splenectomy is performed with utmost care and only when clearly indicated. Regular follow-up assessments are done following splenectomy.

The present study was planned in Department of General surgery, Government Doon Medical College, Dehradun, Uttarakhand, India. The study was conducted from March 2018 to June 2019. In the present study 20 patients diagnosed with the thalassemia in which the splenectomy was suggested. A single team were operated all the patients which headed by the second author. Hypersplenism, increased requirement of blood transfusion, haemosiderosis and abdominal discomfort due to splenomegaly revealed as indications for splenectomy. Preoperative blood transfusion were done to correct Hb >10 gm% in all patients. Immunization was done 3 weeks prior to surgery with polyvalent pneumococcal, hepatitis B and H. influenzae B vaccines for all patients.

The data generated from the present study concludes that Laparoscopic splenectomy revealed to be feasible and safe procedure to β thalassemia patients with splenomegaly. Pfannenstiel incision for specimen extraction is significantly decrease time, with low morbidity and offer better cosmetic results. The operative time in laparoscopic splenectomy is still longer than the open splenectomy but it decrease with time due to increased experience.

Keywords: Laparoscopic splenectomy, Thalassemia, Pfannenstiel incision, etc.

Introduction

The spleen, originally called the organum plenum mysterii by Galen, has long been an important organ for surgeons. The first splenectomy was performed by Andirano Zaccarello in 1549 on a young woman with an enlarged spleen who survived for 6 years after surgery. [1] Traditionally, surgical removal of the spleen was done via an open approach using either an upper midline or left subcostal incision. With the advent of minimally invasive techniques, laparoscopic splenectomy has become a standard procedure for elective removal of the spleen for most indications. Since the first report of laparoscopic splenectomy by Delaitre and Maignien in 1991, [2] it has been increasingly used; however, several technical challenges remain related to removing this fragile, well-vascularized organ that lies close to the stomach, colon, pancreas, and kidney.

Indications for laparoscopic splenectomy are the same as those for open splenectomy except when emergency

splenectomy and exploratory laparotomy for traumatic injuries are needed. Laparoscopic splenectomy is indicated for various benign hematologic diseases, malignant hematologic diseases, secondary hypersplenism, and other anatomic disorders of the spleen. The most common benign hematologic disease treated with laparoscopic splenectomy is immune thrombocytopenic purpura (ITP), and the procedure is recommended when medical therapy, including steroids and intravenous gammaglobulin, fails or long-term steroids are needed. Laparoscopic splenectomy can also be warranted in other benign conditions, including other types of thrombotic purpura, hereditary spherocytosis, major and intermediate thalassemia with secondary hypersplenism or severe anemia, sickle cell disease, and refractory autoimmune hemolytic anemia.

Laparoscopic splenectomy for malignant diseases of the spleen can be performed for diagnostic or therapeutic reasons. Indications include myeloproliferative disorders, lymphoproliferative diseases, hairy cell leukemia,

Hodgkin and non-Hodgkin lymphoma, malignant vascular tumors, malignant lymphomas, and lymphangiosarcomas. Although the use of laparoscopic splenectomy in trauma has been reported, its role has been limited because most hemodynamically stable patients with splenic injuries are successfully treated nonoperatively, and unstable patients require emergency laparotomy for control of hemorrhage and to evaluate possible associated traumatic injuries. However, there is growing evidence that it can be a feasible option in the trauma setting in appropriately selected patients. [3]

Contraindications for laparoscopic splenectomy are similar to those for all laparoscopic surgical procedures. They include the inability to tolerate general anesthesia, uncontrollable coagulopathy, and the need for laparotomy for associated procedures. Although reports on the safety of laparoscopic splenectomy in patients with cirrhosis and portal hypertension have been published, many consider the presence of these conditions an absolute contraindication for laparoscopic splenectomy.

Massive splenomegaly has been regarded as a relative contraindication; however, the hand-assisted technique may facilitate removal of large spleens in a minimally invasive fashion. Good results are being reported for laparoscopic removal of very large spleens, and it has been suggested that with advances in laparoscopic technology and expertise, laparoscopic splenectomy may become the gold standard operation even for massive spleens and splenic malignancies. [4]

A standard laparoscopic tray is used for a laparoscopic splenectomy, including laparoscopic scissors and atraumatic graspers. Telescopes, including a 30° or 45° 5- or 10-mm laparoscope, are used, depending on the surgeon's preference and need for visualization. Three or four trocars are usually needed; one trocar should be a 12-mm port that can be used for laparoscopic stapler introduction and specimen removal. Electrosurgical devices such as an electrothermal bipolar sealing device (Ligasure; Covidien, Mansfield, MA) or ultrasonic coagulation shears (Harmonic; Ethicon Endo-Surgery, Cincinnati, OH) can be used to assist with splenic mobilization and dissection. Typically, the splenic hilar vasculature is divided by using an endoscopic stapling device with a vascular load; however, reports have described the safety and efficacy of the electrothermal bipolar sealing device. [5]

An impervious retrieval bag is needed for morcellation and removal of the specimen. Usually, sacs made from ripcord nylon are used because materials such as

polyurethane are vulnerable to perforation. Special items such as a suction/irrigator and fan or snake retractors for elevation of the spleen are sometimes needed; however, this is left to the discretion of the surgeon. Ring forceps are useful for morcellating the spleen in the retrieval sac.

In the performance of a laparoscopic splenectomy, it is essential always to be mindful that conversion to open surgery may be warranted, possibly on an emergency basis. To prepare for this possibility, when the patient is placed in the lateral position, a wide field should be prepared to allow access to the midline in the event that upper-midline or hand-assist access is needed. Also, it may be helpful to mark the skin two fingerbreadths below the left costal margin before insufflation, in the event that a left subcostal incision proves necessary.

Before splenic mobilization is initiated, diagnostic laparoscopy should be used to look for accessory spleens, which are present in 12-16% of patients and as many as 32% of patients with immune (idiopathic) thrombocytopenia purpura (ITP). Accessory spleens are commonly found in the splenic hilum, along the splenic vessels, in the greater omentum, and in the splenorenal ligament. For large spleens and early in the experience of surgeons undertaking laparoscopic splenectomy, a hand-assisted technique may reduce conversion rates and operating time. Single-port approaches to laparoscopic splenectomy have been described that appear to be safe and effective; however, they have not been shown to have clear advantages over conventional approaches. Single-port techniques will not be described further here. [6]

For the lateral approach, the operation begins with safe laparoscopic abdominal access. This can be accomplished with an open or a closed technique, in accordance with the skill, experience, and comfort level of the surgeon. Although an open cutdown technique for the direct insertion of the first trocar is sometimes favored, an optical trocar technique with preinsufflation using a Veress needle can be quite useful, especially in patients who are obese. The use of the Veress needle is contraindicated in patients with massive splenomegaly or severe thrombocytopenia and in children because of the limited working space and risk of splenic injury and bleeding.

The first trocar, either a 5- or a 12-mm port, is usually placed in the midclavicular line 2-6 cm below the costal margin, depending on the size of the spleen. Preoperative imaging with computed tomography (CT) or ultrasonography (US) can facilitate operative planning by assessing splenic size, locating accessory spleens, and

aiding in decisions regarding port placement and surgical technique (laparoscopic, hand-assisted, or open).

Subsequent trocars are placed after diagnostic laparoscopy; placement varies, depending on the patient's body habitus and spleen size. All ports should be placed 3-4 cm below the inferior tip of the spleen to allow adequate working space for visualization and safe instrument exchange. A medial trocar is placed just off the midline/subxiphoid region in the left subcostal position. A third trocar is placed in the anterior axillary line in the left subcostal region. Bleeding is one of the most common and feared complications related to laparoscopic splenectomy and is the most common reason for conversion to an open approach. [7]

Postoperative bleeding following laparoscopic splenectomy occurs in approximately 3% of patients. Bleeding can be encountered following technical misadventures such as tearing the splenic capsule or failure to adequately control the splenic hilar vessels. Meticulous dissection around the splenic capsule can limit parenchymal tears. The surgeon should try to avoid grasping the spleen. Staplers, clips, and electrosurgical devices can be used to control the splenic hilar vessels, although the surgeon should be prepared if these instruments fail. Subsequent bleeding can usually be controlled with additional clips or ligation of the vessels more proximal, if adequate dissection of the hilar vessels has been carried out. Treatment of postoperative bleeding from the staple line is more challenging and may require a return to the operating room, though the use of postoperative splenic artery embolization has been described with success in one patient. [8]

Pancreatic tail injury is another feared complication of both open and laparoscopic splenectomy, which can cause pancreatic abscesses or fistulas. Careful dissection of the splenic hilum and adequate visualization of the pancreatic tails are mandatory before vessel ligation. It is generally believed that the lateral approach makes this dissection easier and the plane between the pancreatic tail and splenic hilum more visible than is the case with the anterior approach. Drains are rarely necessary in laparoscopic splenectomy; however, if the surgeon is concerned about a possible pancreatic tail injury, a closed suction drain should be used.

Overwhelming postsplenectomy infection (OPSI) is a well-known major long-term risk for splenectomy patients. Patients are at lifelong risk for the development of OPSI; however, the highest risk is in the first 2 years after surgery. Although the reported risk of OPSI is relatively low (3.2%), associated mortalities as high as 40-50% have been described. [9]

Therefore, in patients undergoing elective splenectomy, vaccination against meningococcal, pneumococcal, and Haemophilus influenzae type B infections at least 15 days before the procedure is recommended. In patients undergoing emergency splenectomy, vaccination is recommended within 30 days after the procedure. The pneumococcal vaccine should be repeated every 5 years, and patients should receive an influenza vaccine annually. Accessory spleens are present in as many as 12-32% of patients, and a thorough evaluation for accessory spleens should be made after initial trocar placement. Accessory spleens are typically located in splenic hilum, along the splenic vessels, in the greater omentum, and along the splenorenal ligament and are usually accessible in both the lateral and anterior approach. [10]

Although the risk of missing accessory spleens was once a proposed shortcoming of the laparoscopic approach, the detection rates for accessory spleens with laparoscopy appear to be similar to those with the open approach. [11]

Portal vein thrombosis is increasingly being recognized as a complication of splenectomy and should be considered in patients suffering from postoperative anorexia, abdominal pain, ileus, low-grade fevers, and elevated platelet and leukocyte counts. Portal vein thrombosis has been reported to occur in 0.7-14% of patients. Risk factors associated with portal vein thrombosis include splenomegaly, myeloproliferative disorders, and hemolysis, with incidences reported to be as high as 80% in these high-risk patients. [11] In one study, a platelet count increasing to more than eight times the baseline preoperative level after surgery was a risk factor for portal vein thrombosis after laparoscopic splenectomy. [12] A preoperative splenic vein diameter of 8 mm or greater has also been suggested as a risk factor for portal or splenic vein thrombosis. Whether the technique of surgery (ie, open or laparoscopic) affects the rate of portal vein thrombosis remains unclear. Anticoagulation therapy is recommended for all symptomatic patients.

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utmost care and only when clearly indicated. Regular follow-up assessments are done following splenectomy.

Methodology:

The present study was planned in Department of General surgery, Government Doon Medical College, Dehradun, Uttarakhand, India. The study was conducted from March 2018 to June 2019. In the present study 20 patients diagnosed with the thalassemia in which the splenectomy was suggested. A single team were operated all the patients which headed by the second author. Hypersplenism, increased requirement of blood transfusion, haemosiderosis and abdominal discomfort due to splenomegaly revealed as indications for splenectomy. Preoperative blood transfusion were done to correct Hb >10 gm% in all patients. Immunization was done 3 weeks prior to surgery with polyvalent pneumococcal, hepatitis B and H. influenzae B vaccines for all patients.

All the patients were informed consents. The aim and the objective of the present study were conveyed to them. Approval of the institutional ethical committee was taken prior to conduct of this study.

Results & Discussion:

The use of splenectomy in thalassemia has declined in recent years. This is partly due to a decreased prevalence of hypersplenism in adequately transfused patients. There is also an increased appreciation of the adverse effects of splenectomy on blood coagulation. In general, splenectomy should be avoided unless absolutely indicated.

Splenectomy is indicated in the transfusion-dependent patient when hypersplenism increases blood transfusion requirement and prevents adequate control of body iron with chelation therapy. An enlarged spleen—without an associated increase in transfusion requirement—is not necessarily an indication for surgery. Patients with hypersplenism may have moderate to enormous splenomegaly, and some degree of neutropenia or thrombocytopenia may be present.

Annual transfusion volume exceeding 225 to 250 mL/kg per year with packed red blood cells (hematocrit 75 percent) may indicate the presence of hypersplenism. The volume calculation should be corrected if the average hematocrit is less than 75 percent. The possible development of alloantibody should also be ruled out. Splenectomy should be avoided unless there is an inability to maintain iron balance with optimal chelation, or if there are clinically significant complications such as

pancytopenia and marked enlargement. Often, hypersplenism develops because of a low pre-transfusion hemoglobin. Increasing the pre-transfusion hemoglobin to between 9.5 and 10 may reverse hypersplenism.

β -thalassemias are a group of hereditary blood disorders characterized by anomalies in the synthesis of the beta chains of Hb, resulting in variable phenotypes ranging from severe anemia to clinically asymptomatic individuals. [14] The worldwide annual incidence of symptomatic individuals is estimated to be 1/1,00,000 population. Though categorized as a haematological disorder, it causes multisystem involvement damaging the heart, liver, bones, kidneys and many endocrine glands; leading to significant morbidity in these children. [14-17]

Table 1: Basic Details:

Parameters	No. of Cases
Age:	
5 – 10 years	0
10 – 15 years	12
15 – 20 years	8
Sex:	
Male	13
Females	7
Total Cases	20
Spleen Diameters cm	10.2 – 20.5

Table 2: Intraoperative Observations

Intraoperative Observations	Observations
Intra-operative blood loss (ml)	142 – 215
Operation time (min)	85 – 126
Intraoperative blood transfusion	1 Case
Intraoperative haemorrhage	1 Case
Conversion to open	1 Case
Splenic weight (gm)	875 - 1195

Table 3: Postoperative Observations

Postoperative Observations	Observations
Time to start oral (hrs)	27.1 – 35.3
Port site infection	1 Case
Intra-abdominal haemorrhage	1 Case
Ileus and diarrhoea	0
Duration of hospital stay (days)	1.5 – 3.5

Various authors like Machado et al., al-Salem et al., al-Hawsawi et al. have previously published series with splenectomy in thalassemia patients and the benefits of splenectomy have been found to be unequivocal by one and all. [18-20] Various series by Targarona et al., Wolff et al. have detected decreased PRBC transfusion requirement and less drop in Hb following splenectomy. [20-21] However, none of the previously published studies have taken into account and objectively assessed

the impact of splenectomy on overall quality of life of children with β -thalassemia.

Laparoscopic splenectomy was first described in 1991 by Delaitre and Maignien. [23] In 1993 it was first reported in children by Lobe, et al. Since then significant improvements in instrumentation and technology such as harmonic scalpel, endovascular staplers have increased the ease of performance and scope of this type of surgery.

A combined review of 5 published series of open splenectomy in 611 patients revealed morbidity in 26% and mortality in 3.1%. A combined review of 461 cases from 15 published series revealed morbidity in 8%, mortality in 0.65% and conversion to open surgery in 8%. Katkhouda reported the largest series of 103 laparoscopic splenectomies in 1998. [24] They did not have any mortality, requiring conversion to open surgery in 4 patients and complications in 6 patients. Harold et al in 1999 reported a mean hospital stay of 1.5 days after laparoscopic splenectomy. [25] Friedman et al reported their experience of 63 laparoscopic splenectomies in 1997. Only 3 patients required blood transfusion during surgery. [26] With the current published data and increased experience, laparoscopic splenectomy is emerging as a gold standard for the management of haematological disorders.

Preoperative embolization of splenic artery could be used to decrease the blood loss and decrease the operative time for surgery of large spleens, but because of the serious complications, as severe pain, perisplenitis and abscess of the spleen, this procedure became not advised. [27] So did not follow preoperative embolization in the patients was not followed.

Conclusion:

The data generated from the present study concludes that Laparoscopic splenectomy revealed to be feasible and safe procedure to β thalassemia patients with splenomegaly. Pfannenstiell incision for specimen extraction is significantly decrease time, with low morbidity and offer better cosmetic results. The operative time in laparoscopic splenectomy is still longer than the open splenectomy but it decrease with time due to increased experience.

References:

1. Coon WW. The spleen and splenectomy. *Surg Gynecol Obstet.* 1991 Nov. 173 (5):407-14.
2. Delaitre B, Maignien B. [Splenectomy by the laparoscopic approach. Report of a case]. *Presse Med.* 1991 Dec 21-28. 20 (44):2263.
3. Shamim AA, Zafar SN, Nizam W, Zeineddin A, Ortega G, Fullum TM, et al. Laparoscopic Splenectomy for Trauma. *JLS.* 2018 Oct-Dec. 22 (4).
4. Somasundaram SK, Massey L, Gooch D, Reed J, Menzies D. Laparoscopic splenectomy is emerging 'gold standard' treatment even for massive spleens. *Ann R Coll Surg Engl.* 2015 Jul. 97 (5):345-8.
5. Misawa T, Yoshida K, Iida T, Sakamoto T, Gocho T, Hirohara S, et al. Minimizing intraoperative bleeding using a vessel-sealing system and splenic hilum hanging maneuver in laparoscopic splenectomy. *J Hepatobiliary Pancreat Surg.* 2009. 16 (6):786-91.
6. Wu S, Lai H, Zhao J, Deng X, Wei J, Liang J, et al. Systematic review and meta-analysis of single-incision versus conventional multiport laparoscopic splenectomy. *J Minim Access Surg.* 2018 Jan-Mar. 14 (1):1-8.
7. Glasgow RE, Mulvihill SJ. Laparoscopic splenectomy. *World J Surg.* 1999 Apr. 23 (4):384-8.
8. Kercher KW, Novitsky YW, Czerniach DR, Litwin DE. Staple line bleeding following laparoscopic splenectomy: intraoperative prevention and postoperative management with splenic artery embolization. *Surg Laparosc Endosc Percutan Tech.* 2003 Oct. 13 (5):353-6.
9. [Guideline] Habermalz B, Sauerland S, Decker G, Delaitre B, Gigot JF, Leandros E, et al. Laparoscopic splenectomy: the clinical practice guidelines of the European Association for Endoscopic Surgery (EAES). *Surg Endosc.* 2008 Apr. 22 (4):821-48.
10. Mortelé KJ, Mortelé B, Silverman SG. CT features of the accessory spleen. *AJR Am J Roentgenol.* 2004 Dec. 183 (6):1653-7.
11. Romano F, Caprotti R, Conti M, Piacentini MG, Uggeri F, Motta V, et al. Thrombosis of the splenoportal axis after splenectomy. *Langenbecks Arch Surg.* 2006 Sep. 391 (5):483-8.
12. Huang D, Tao M, Cao L, Wang X, Zheng S, Cao Y. Risk Factors and Anticoagulation Effects of Portal Vein System Thrombosis After Laparoscopic Splenectomy in Patients With or Without Cirrhosis. *Surg Laparosc Endosc Percutan Tech.* 2019 Dec. 29 (6):498-502.
13. Colah RB, Gorakshakar AC, Nadkarni AH. Invasive & non-invasive approaches for prenatal diagnosis of haemoglobinopathies: Experiences from India. *Indian J Med Res* 2011;134:552-60.
14. Galanello R, Origa R. Beta-thalassemia. *Orphanet J Rare Dis* 2010;5:11.
15. Borgna-Pignatti C, Cappellini MD, De Stefano P, Del Vecchio GC, Forni GL, Gamberini MR, et al. Survival and complications in thalassemia. *Ann N Y Acad Sci* 2005;1054:40-7.
16. DeBaun MR, Frei-Jones M, Vinchinsky E. Thalassemia syndromes. In: Kliegman RM, Stanton BF, St Geme JW, Schor NF, Behrman RE, editors. *Nelson Textbook of Pediatrics.* 19th ed. Philadelphia: Elsevier; 2008. p. 1674-7.
17. Schrier SL, Angelucci E. New strategies in the treatment of the thalassemias. *Annu Rev Med* 2005;56:157-71.
18. al-Salem AH, al-Dabbous I, Bhamidibati P. The role of partial splenectomy in children with thalassemia. *Eur J Pediatr Surg* 1998;8:334-8.
19. Al-Salem AH, Nasserulla Z. Splenectomy for children with thalassemia. *Int Surg* 2002;87:269-73.
20. al Hawsawi ZM, Hummaida TI, Ismail GA. Splenectomy in thalassaemia major: experience at Madina Maternity and Children's Hospital, Saudi Arabia. *Ann Trop Paediatr* 2001;21:155-8.
21. Targarona EM, Espert JJ, Cerdán G, Balagué C, Piulachs J, Sugañes G, et al. Effect of spleen size on splenectomy outcome. A comparison of open and laparoscopic surgery. *Surg Endosc* 1999;13:559-62.
22. Wolff JA, Sitarz AL, Vonhofe FH. Effect of splenectomy on thalassemia. *Pediatrics* 1960;26:674-8.

23. Katkhouda N, Mavor E. Laparoscopic Splenectomy. Surg Clin North Am 2000; 80 (4) : 1285-97.
24. Katkhouda N, Hurwitz MB, Rivera RT, et al. Laparoscopic splenectomy: Outcome and efficacy in 103 consecutive patients. Ann Surg 1998; 228 : 568.
25. Harold KL, Schlinkert RT, Mann DK, et al. Long-term results of laparoscopic splenectomy for immune thrombocytopenic purpura. Mayo Clin Proc 1999; 74: 37.
26. Friedman RL, Hiatt JR, Korman JL, et al. Laparoscopic or open splenectomy for hematologic disease: Which approach is superior? J Am Coll Surg 1997; 185 : 49.
27. Poulin EC, Thibault C, Mamazza J. Laparoscopic splenectomy. Surg Endo. 1995;9(2):172-7.