

Laparoscopic splenectomy in patients under the age of eighteen. Experience in 18 cases

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SUMMARY: Laparoscopic splenectomy in patients under the age of eighteen. Experience in 18 cases

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Background. Pediatric surgery is now in the forefront of minimal access procedures. Although pediatric surgeons have been skeptic about laparoscopic splenectomy, recently minimally invasive approach for spleen removal has been reevaluated also in young patients. Purpose of this study was to report Authors' personal experience in patients under eighteen who underwent laparoscopic splenectomy. Results of the procedure were evaluated.

Patients and methods. A retrospective review was undertaken in 18 splenectomised patients under the age of eighteen. Indications were hereditary spherocytosis in 10 patients, β -thalassemia in 4, idiopathic thrombocytopenic purpura in 3 and a splenic cyst in 1 child.

Results. No intra-operative complications occurred. No conversion to open surgery was reported. During the follow-up one case of portal vein thrombosis, treated by medical therapy, was encountered and no other postoperative complications were observed.

Conclusions. Laparoscopic approach has to be preferable for all those children undergoing spleen surgery. In experienced hands, it is of beneficial effects with a very reasonable rate of complications.

RIASSUNTO: Splenectomia laparoscopica in pazienti di età inferiore a 18 anni. Esperienza personale su 18 casi.

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Premessa. La chirurgia pediatrica è ormai all'avanguardia con le procedure mini-invasive. Sebbene in passato i chirurghi pediatri abbiano dimostrato scetticismo sulla splenectomia laparoscopica, recentemente l'approccio mini-invasivo per l'asportazione della milza è stato rivalutato anche nei pazienti più giovani. Lo scopo dello studio è quello di riportare l'esperienza personale degli Autori in pazienti di età inferiore a 18 anni splenectomizzati per via laparoscopica e valutarne i risultati.

Pazienti e metodi. Uno studio retrospettivo è stato effettuato su 18 pazienti di età inferiore a 18 anni sottoposti a intervento di splenectomia laparoscopica totale. Le indicazioni al trattamento chirurgico sono state: sferocitosi ereditaria in 10 pazienti, β -talassemia in 4, porpora trombocitopenica idiopatica in 3 ed un singolo caso di cisti splenica.

Risultati. Non si è verificata alcuna complicanza intraoperatoria. Non è stata necessaria alcuna conversione. Durante il follow-up si è manifestato un solo caso di trombosi della vena porta, trattato peraltro con terapia medica.

Conclusioni. L'approccio laparoscopico è sempre da preferirsi per i bambini che richiedono la chirurgia splenica. In mani esperte si è dimostrato di gran beneficio, associato da un esiguo numero di complicanze.

KEY WORDS: Laparoscopic splenectomy - Pediatric surgery - Spleen diseases.
Splenectomia laparoscopica - Chirurgia pediatrica - Malattie spleniche.

Introduction

Since 1991, when it was first performed (1), laparoscopic splenectomy has been increasingly accepted and

has become the technique of choice for surgical removal of the spleen in referral centers. Minimally invasive approach has been preferred for splenectomy not only for cosmetic reasons but also for immunologic benefits, cost advantages and reasonable rate of complications.

However, pediatric surgeons have been slower to adopt minimal-access surgery of the spleen since traditional operations in children are already performed with minimal incisions and patients are often too small for the feasibility of laparoscopic approach. Moreover, many of the conditions that require spleen surgery in pediatric age are rare (2,3).

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But as time goes on, new surgical miniaturized instruments, which can be inserted in abdomen through a few millimeter hole, and increased experience of surgical teams allowed to use this technique in children with low risk (4).

The most frequent conditions requiring spleen surgery both in adults and children are hematologic and immunologic disorders, and trauma. Most common indications for splenectomy in children are hereditary spherocytosis and idiopathic thrombocytopenic purpura. Other indications are spleen trauma, thalassemia and sickle cell disease. On the other side hemoglobin H, Coomb's anemia, staging in Hodgkin's disease, leukemia, Gaucher's disease, and portal hypertension are uncommon. Splenectomy is reserved, anyway, for patients with hematologic and immunologic disease in whom medical therapy has failed. In fact, the importance of preserving the spleen in order to maintain the host's immunologic response has been widely recognized, especially in pediatric age, and therefore, nonoperative therapy is the first-line management for almost all splenic conditions (5).

In this study Authors' initial experience in eighteen patients under eighteen years of age who underwent laparoscopic splenectomy is reported. Indications, technique and results of laparoscopic spleen surgery in pediatric age is discussed and the advantage of minimally invasive approach is emphasized.

Patients and methods

From January 1998 to September 2009, the charts of eighteen patients under the age of eighteen who underwent laparoscopic total splenectomy (out of 162 splenectomies of all ages) were retrospectively analyzed. The group consisted of 8 boys and 10 girls, aged 8 to 17 years with a mean age of 12 years. Indications for splenectomy were hereditary spherocytosis (HS) in 10 patients, β -thalassemia (β -thal) in 4, idiopathic thrombocytopenic purpura (ITP) in 3 and a splenic cyst in 1 child.

In patients with symptomatic ITP, preoperative platelet count was under 20,000 per mm^3 . Children with hemolytic disorders (HS and β -thal) underwent preoperative ultrasound in order to search for cholelithiasis and two out of ten of those affected by HS showed stones in the gallbladder. All children received preoperative immunizations with pneumococcal vaccine, meningococcal group C vaccine, and Haemophilus influenzae type B vaccine according to guidelines for prevention of overwhelming postsplenectomy sepsis. An antibiotic prophylaxis was administered to all children in the evening before surgery. The antibiotic of choice was a single shot of intramuscular benzylpenicillin. Furthermore suicide inhibitor piperacillin/tazobactam's first dose was given one hour before splenectomy and continued for other three days after surgery.

Laparoscopic splenectomies were performed with patients lying in the right lateral position (left flank elevated about 30° above the operating table). An Hasson trocar was inserted in the upper umbilical region, and three other trocars were placed in the subxyphoid area (5 mm), lower left midclavicular line below the umbilical line (10 mm), and lower left axillary line (5 mm). Like in the open approach, during the initial part of the procedure, sites of accessory spleens were accurately scoped. With a 30° scope, after dissection of the sple-

no-colic ligament so as to mobilize the lower pole, division of the gastrosplenic ligament with the short gastric vessels was accomplished by using the ultrasonic dissector (Ultracision Harmonic Scalpel, Ethicon Endosurgery, Inc., Cincinnati, OH, USA) (Fig. 1). At that moment splenic artery was recognized, mobilized and double clipped, in order to avoid bleedings during the following steps of the procedure (Fig. 2). The convex surface of the spleen was then accurately dissected. At that point, hanging the spleen, the splenic hilum was approached from the anterior aspect. Splenic artery and vein were clipped and divided, taking care not to damage pancreatic tail, using a stapler device (Endo-GIA Vascular Stapler, Ethicon Endosurgery, Inc., Cincinnati, OH, USA) (Fig. 3). The spleen was then inserted in a plastic bag (endobag) that was pulled through the abdominal umbilical access. It was then crushed and removed, piece by piece, trying to avoid bag lacerations and splenic fragments' peritoneal implantation (splenosis).

After splenectomy all patients continued antibiotic prophylaxis with piperacillin/tazobactam as above-mentioned for a total period of 4 days followed by amoxicillin for 3 weeks.

Results

Laparoscopic splenectomies' data and results are reported in table 1. The mean operative time for laparoscopic splenectomy in Authors' experience was 50 minutes (range 40 to 75). In all analyzed children intraoperative complications were not encountered and conversion to open surgery was not necessary in any patient. In two out of ten cases of HS, that showed gallstones during US preoperative evaluation, a concomitant cholecystectomy was performed. Accessory spleens were detected and removed during surgery in two patients with HS. Mean hospital stay was 4 days (range 3 to 7). Patients were followed for a period of time ranging from 3 months to 2 years (mean follow-up time 16 months). During this follow-up no infective complications occurred

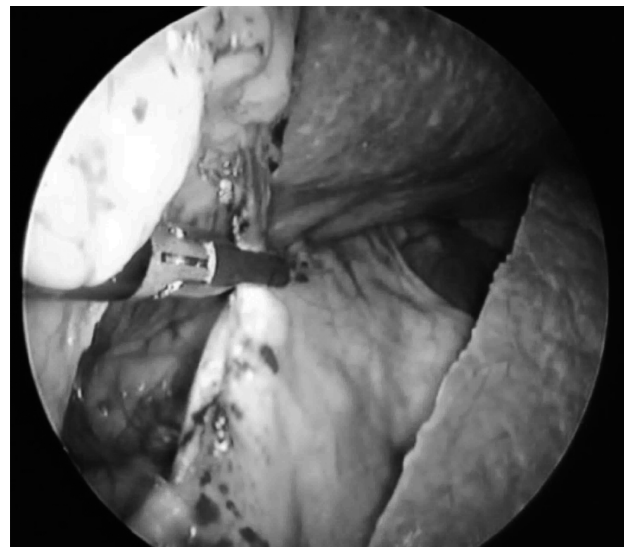


Fig. 1 - Section of gastro-splenic ligament with short gastric vessels.

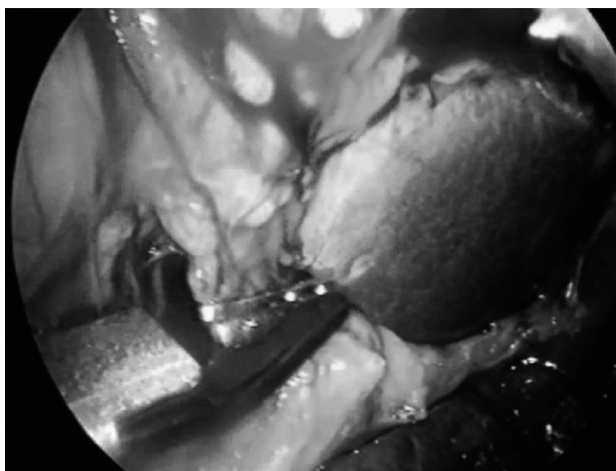


Fig. 2 - Occlusion of splenic artery by clips application.



Fig. 3 - Dissection of the splenic hilum with splenic artery and vein by stapler device.

and the only postoperative complication was one case of portal vein thrombosis successfully treated with medical therapy (heparin). All children affected by HS and β -thal reduced significantly the need for transfusion. Two out of three ITP cases shows resolution of symptoms related to thrombocytopenia and an increasing platelet count up to 80,000 per mm^3 six months after splenectomy.

Discussion

The first laparoscopic splenectomy in pediatric age was performed by Tulman and Holcomb in 1993.⁶ For years, however, peritoneoscopy in children was limited mainly to diagnostic purpose. This might be partly due to the uncertainty about potential advantage of “keyho-

le” surgery in children. Since small patients have already small incisions in open surgery, several surgeons felt that the benefit of minimally invasive surgery in children was less evident. Nonetheless, surgery is traumatic anyway, and initially small wound could become a much bigger and rather unsightly scar in later life.

For these reasons a few practitioners saw an opportunity to minimize the insult of operation in their young patients by the minimally invasive surgery increasing thereby cosmetic satisfaction. Furthermore, also in children, as it has been reported in adults, smaller incisions are responsible for less post-traumatic pain which results in a shorter duration of postoperative ileus, less analgesic requirement, less wound complications, minimal immunosuppression, shorter hospital stays and, last but not least, less expensive hospital-stay associated costs. In addition, youngsters return to full activity quickly and an earlier return to preoperative schooling activities for children may actually free parents and allow their earlier return to normal work and productivity.

Minimal access surgery is of huge advantage also for surgeons. The great visual details obtained thanks to the magnification of the optic system in laparoscopic surgery allow a better knowledge and recognition of the human anatomy. It results also in a complete detailed exploration of the abdomen with the further possibility to diagnose associated pathologies. The aphorism: “Surgery is to watch” has finally found a solution (4, 7, 8).

The most common indications for total splenectomy in children are hematologic disorders. They include hereditary spherocytosis, idiopathic thrombocytopenic purpura, β -thalassemia and sickle cell disease. In Authors' experience HS was indication for surgery in 56% of the group's patients, confirming data from literature where similar results are reported (9-12). In this disease the fundamental abnormality is an erythrocyte membrane loss which is associated with defective proteins, in particular spectrin and ankyrin, and results in an abnormal spherical red cell shape. This is responsible for red cells to be less deformable and more susceptible to trapping and consequent disintegration within the spleen's sinuses. Removing splenic tissue decreases premature destruction of fragile red blood cells (13, 14). Total splenectomy in HS has been shown to reduce hemolysis, thus prolonging the red cell life span and improving severe anemia. In addition, splenectomy has also been shown to decrease serum bilirubin thereby reducing pigmented gallstones formation. After splenectomy patients with HS decrease blood transfusion requirement, like it happened in all ten cases of the study (10, 15).

Laparoscopic surgery in patients where removal of the spleen is indicated for HS or ITP could be of huge value also for detection of accessory spleens. They are thought to be present in 10 to 25% of the population.¹⁶ Although accessory spleens frequently are located near

TABLE 1 - PERSONAL EXPERIENCE IN PATIENTS UNDER 18 YEARS WHO UNDERWENT LAPAROSCOPIC SPLENECTOMY.

Period of study	Jan 1998 - Sep 2009
Laparoscopic splenectomies < 18 yrs	18 (out of 162 splenectomies)
Male/Female	8/10
Mean age (range)	12 yrs (8 - 17)
Indications:	
Hereditary Spherocytosis (HS)	10 (56%)
β -thalassemia (β -thal)	4 (22%)
Idiopathic Thrombocytopenic Purpura (ITP)	3 (17%)
Splenic cyst	1 (5%)
Mean operative time (range)	50 min (40 - 75)
Concomitant cholecystectomy	2 HS pts
Accessory spleens	2 HS pts
Intraoperative complications	0
Conversion to open surgery	0
Mean hospital stay (range)	4 days (3 - 7)
Mean follow-up time (range)	16 mths (3 - 24)
Postoperative complications	1 portal vein thrombosis
Infective complications	0

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the splenic hilum and on the left upper quadrant, they may very rarely have intrapancreatic, intrahepatic and pelvic locations (17). For accessory spleens, preoperative or perioperative detection and immediate removal during the operation is the main aim of the surgeons to achieve a successful result. Among patients who initially seem to respond to splenectomy 25% will relapse. Up to 25% of these recurrent cases might have accessory spleens, missed and not removed during splenectomy (18).

Since preoperative imaging modalities still have limited sensitivity (less than 80%) (17), surgery is essential for discovery of accessory spleens. Although some study do not reveal any significant difference between laparoscopic and conventional splenectomy in assessing intraoperatively the presence and location of accessory spleens (19, 20), it is in the opinion of the Authors that laparoscopic surgery, when compared to open traditional surgery, allows a better visualization of the anatomic regions where accessory spleens are usually located.

In order to search for accessory spleens, surgeon should start the operation with the full exploration of the abdomen. This primary maneuver is important, since if it is applied at the end of the intervention it can be more difficult, especially if small hematomas and blood clots lie in the operative field. Intraoperative ultrasonography is also recommended but accuracy of this technique has not been established yet (21).

Another indication for laparoscopic splenectomy, which is commonly reported in literature, is β -thalassemia,

usually in patients with massive splenomegaly. In the reported Authors' experience, β -thalassemia accounted for 22% of cases. It has been debated in literature if, in case of splenomegaly, splenectomy should be performed laparoscopically or by open surgery (22, 23). Some Author reported a significant difference concerning conversion rate to open surgery in patients who underwent laparoscopic splenectomy whereas mean spleen longitudinal diameter was superior to 28.4 cm compared to patients with a normal spleen volume or a mild splenomegaly (24).

According to the Authors of this report, with increasing experience, splenomegaly could be managed by laparoscopic surgeons without any additional risk. The Authors of the present study performed laparoscopic splenectomy in adult patients even when the spleen is larger than 30 cm in longitudinal diameter, according to other Authors who agree that splenomegaly should not be considered a contraindication for laparoscopic splenectomy (22,23).

In ITP patients laparoscopy has a preminent role when removal of the spleen is indicated. Response to splenectomy in patients where medical therapy with corticosteroids has failed, can be predictive in selected cases (25). Laparoscopy in these indications has been considered the gold standard for spleen removal probably because of the less challenging operative technique due to the absence or mild form of splenomegaly in these patients. Spleen size is indeed usually normal in case of ITP. Moreover laparoscopic approach, as above-mentioned,

allows complete visualization of sites where accessory spleens are located, whose detection and retrieval is essential for the final positive result (26).

The benefits of laparoscopic splenectomy must be balanced against the immediate and long-term risks of the procedure. Surgeons have to face with intraoperative complications such as internal hemorrhage and parenchymal injuries like pancreatic lesions, diaphragm and bowel perforations. The incidence of technical complications usually dropped dramatically with increasing experience in laparoscopic surgery (4, 11, 28). The technique above reported by the Authors can reduce, if properly accomplished, the risks while performing the procedure. The employment of the open Hasson method for the access in the abdominal cavity is associated with a decreased risk of perforation or damage to the intra-abdominal organs (4). The lateral position of the patient is in Authors' opinion essential for a good exposure of the short gastric vessels and of the splenic hilum, therefore helping to prevent bleeding related to their accidental damage. Intraoperative hemorrhage can also be minimized if preventive occlusion of the splenic artery is accomplished. Pancreatic injuries can be avoided if the splenic hilum is sectioned after complete mobilization of the spleen and hanging maneuver of the organ is performed (27).

With the above mentioned technical pearls, with a proper training and increase in experience, laparoscopic surgery can be safely performed in young infants and even small neonates with minimal morbidity and mortality (4,29).

Among postoperative complications overwhelming postsplenectomy sepsis (OPSS) and portal vein thrombosis are considered the most life-threatening. Postsplenectomy sepsis is usually fulminant and occurs days to years after removal of the spleen. The risk of OPSS is highest in children, especially those under two years of age and during the first two years after splenectomy. There are, however, reported cases of fulminant sepsis 20 to 40 years after splenectomy, indicating that post-splenectomy patients carry a lifelong risk.

The risk of OPSS' incidence can also be stratified by underlying disease. The lowest risk is related to idiopathic thrombocytopenic purpura followed by sickle hemoglobin disease and trauma; intermediate risk to spherocytosis, and highest risk in thalassaemia. The dramatic nature of the illness is further reflected by the time from initial symptoms to death, with 68% of the deaths occurring within 24 hours and 80% within 48 hours (30). The risk in a splenectomised patients occurs in 1 in 100 patients and is at least 10 times higher the risk of overwhelming infection in the general population. The sepsis will be lethal in 50% of the adults and in almost all of the children. Thalassaemia is still the condition with the highest risk (5, 31,32).

Streptococcus pneumoniae is the most common organism involved in OPSS and the causative agent in 50 to 90% of the cases. *Haemophilus influenzae* type B is the second most common organism related to OPSS. *Neisseria meningitidis* has been cited as the third most common cause of OPSS (30).

To avoid OPSS pediatricians and hematologists have developed programs of prophylaxis with antibiotics and vaccines (32). In the opinion of the Authors, all splenectomised patients should receive pneumococcal immunisation. Patients not previously immunised should receive *Haemophilus influenzae* type B vaccine and meningococcal group C conjugate vaccine. Influenza immunisation should be given too (33). Most authorities recommend antibiotic prophylaxis for asplenic children, sometimes for a long period after splenectomy. Patients should be informed about their increased susceptibility to certain infections, the potential seriousness of OPSS and its possible very rapidly progressive and life-threatening course (30). Partial splenectomy, whenever possible, have also been advocated in order to preserve the immune role of the spleen (32).

The occurrence of thrombosis in the portal system is an underappreciated complication of splenectomy. Various investigators described this event as an uncommon or rare complication, although the more extensive use of radiologic imaging now seems to indicate that it probably occurs more often than previously suspected (34).

The actual incidence of post-splenectomy extrahepatic portal system thrombosis is not clearly determined, ranging between 0.7 and 80% (35). Thrombosis can occur even as late as 2-4 months after splenectomy (34,36). Portal vein thrombosis (PVT) may complicate splenectomy in patients with hemolytic anemia and myeloproliferative disease, whereas the frequency of portal vein thrombosis in case of trauma is not defined (37).

Spleen weight is the only significant factor predictive of postoperative thrombosis. The combination of splenomegaly and an elevated preoperative platelet count was associated with a 75% incidence of this complication (34).

Portal thrombosis should be suspected in patients with fever or abdominal pain after splenectomy. Other patients reported atypical symptoms like abdominal pain, ileus, fever or diarrhea.

An aggressive approach to thrombotic prophylaxis should be considered in high risk patients with massive splenomegaly. Selected patients should receive perioperative intravenous therapy with low-molecular-weight heparin followed by low-dose warfarin for the first weeks after splenectomy or routine antiplatelet therapy with aspirin. As a matter of fact, subcutaneous heparin alone seems to be unable to prevent the development of PVT in high risk patients. Eventually PVT requires suspicion, so as to establish prophylaxis, early diagnosis and appropriate treatment (35).

Conclusions

Minimal access surgery is a new different way to do surgery. When feasible, minimally invasive approach should be considered and preferred to the traditional open procedure for all those children undergoing spleen surgery. Advances in technology along with increased la-

paroscopic experience allow several benefits and few risks. Laparoscopic splenectomy is thereby safe and effective in children. Guidelines for prevention of sepsis and portal vein thrombosis must be followed before and after surgery in splenectomised patients. Proper technique and adequate surgical team training and experience are essential to perform the procedure in a safe way.

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