

Laparoscopic partial splenectomy in pediatric patients

Report of meeting held in Rome , 11th December 2015

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Hematological indications of the partial splenectomy in children

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Children with hematological disease ,which can have beneficial from a partial and/or total splenectomy, may be divided into three major groups. The first group includes the congenital hemolytic anemias in which the spherocytosis plays a significant role. The second group concerns the hemoglobinopathies especially thalassemia major and sickle cell disease (SCD), and the third the acquired immune-hematological disorders such as autoimmune hemolytic anemia and thrombocytopenic purpura. Taking into account the focus of this meeting, the partial splenectomy, it seems appropriate to underline the primary physiological rules of the spleen as an organ of the reticuloendothelial system: the filter function, the hemocatheresis and the bacteria phagocytosis. This last activity, which is part of the immunological function, regards the antigen specific immune response to the production of opsonine, that allows phagocytosis of polysaccharidic bacteria, the main source of sepsis for splenectomized patients. Total splenectomy exposes the patients to a higher risk of sepsis as well the age: children with an age less than five years-old have a 10 % risk of serious infections compared to 1-2% of an adult.

We know from the experience that the underlying disease, such as thalassemia, can increases this risk up to a maximum risk of 1,000 times higher than the risk of 50 of post-trauma splenectomised patient. Looking over hematological indications for splenectomy, these have the main objective to remove the hemocatheresis function of the spleen. The main question is whether the partial splenectomy can preserve the immune function abolishing that of hemocatheresis. A removal of 75% of the splenic volume allows to maintain an immunological response to the pneumococcal stimulus. Hematological and immunological studies have shown that in the patients where partial splenectomy have been performed, the production of immunoglobulins and lymphocytes is maintained with good results. Even in experimental models, when the 75% of the weight of the spleen is removed, they have a preservation of the reticulo-endothelial function, as well as the index of phagocytosis in response to the pneumococcal infection. This feature is not present in the experimental model of auto-transplant splenic tissue due to the vascular modifications that does not allow to preserve the immunological function. Going back to hemocatheresis, we know that an indication to partial splenectomy is spherocytosis, which we classify into different levels of severity according to hemoglobin, reticulocyte and bilirubin values. The splenectomy is reserved for severe patients, which account for approximately 10-15% of all forms of spherocytosis but the objective is to postpone at least till the age of 5-6 years the timing of the surgery. In the long term follow-up

of these patients, Hb values remain stable over time, as well as the reticulocyte count, although the hemolysis is not entirely abolished. If we look in detail to the splenic remnant, we may observe a recurrence of hypersplenism, that still tends to increase over the years, although this does not appear to affect the hematologic parameters of these patients; once again the growth of the spleen, accompanied by a restructuring of the parenchyma and vascular flow seems not to have a significant influence in these patients.

Comparing papers on spherocytosis, the risk of recurrence of hypersplenism is also reported with a small number of patients requiring a total splenectomy. This procedure has been performed with good results also in patient with thalassemia major, although some Authors highlight that this procedure might have only a temporary effect, due to the presence of the residual splenic tissue that can grow in the following years; but at the mean time, this decision could be considered especially at a young age when these children are more sensitive to the infections of capsular bacteria. Considering sickle cell disease and guidelines of the onco-hematology pediatric community, the level of evidence, as well as the indication for the surgery becomes more restrictive; the partial removal of the spleen provides an efficient volume reduction, the need for transfusions and reduces the risk of splenic sequestration. In these patients the clinical picture is more complex as sickle cell disease involves a risk of having splenic infarction; furthermore, despite an appropriate splenic volume we have kids with reduced or functional asplenia where the immunological function is compromised.

A report comparing the partial splenectomy with total laparoscopic to open surgery, shows that there are many, non randomized, prospective studies with heterogeneous data; the hematologic outcome has been analysed showing comparable results with partial and total splenectomy. With regard to the immunological function, the partial splenectomy appears to preserve the activity of phagocytosis. Significant differences were not observed between the open and the laparoscopic approach. In the last group of acquired immunological splenic diseases, the development of immunosuppressive therapies has reduced the indications to the splenectomy. In conclusion, we have still some open questions: partial splenectomy has always an indication or conversely, we must observe some criteria such as a younger age of 5-6 years? Some authors have suggested to reserve this approach to patients that for geographical or logistical reasons do not have a transfusion support and / or a poor antibiotic prophylaxis compliance. A further question to be considered is: the partial splenectomy, preserving the hematological function, could avoid in these patients, the antibiotic prophylaxis and pre-splenectomy vaccinations? Based on the American group' experience, however, antibiotic prophylaxis is recommended for at least one year and still are carried out vaccinations, doubting the long-term effect of the partial splenectomy.

Recommended lectures

The protection of small amounts of splenic tissue against intravenous pneumococcal challenge following subtotal splenectomy (pages 145–149)

Jane F. Goldthorn, Bonnie A. New and Dr. Allen D. Schwartz

Version of Record online: 19 JUL 2006 | DOI: 10.1002/jso.2930170208

Functions of the splenic remnant after subtotal splenectomy for treatment of severe splenic injuries.

Resende V1, Petroianu A. Am J Surg. 2003 Apr;185(4):311-5.

The influence of splenic weight and function on survival after experimental pneumococcal infection

Malangoni MA, Dawes LG, Droege EA, Almagro UA. Ann Surg. 1985 Sep;202(3):323-8.

Hereditary spherocytosis—defects in proteins that connect the membrane skeleton to the lipid bilayer

Eber S1, Lux SE. *Semin Hematol.* 2004 Apr;41(2):118-41.

Long-term evaluation of the beneficial effect of subtotal splenectomy for management of hereditary spherocytosis

Bader-Meunier B, Gauthier F, Archambaud F, Cynober T, Miélot F, Dommergues JP, Warszawski J, Mohandas N, Tchernia G. *Blood.* 2001 Jan 15;97(2):399-403.

Splenectomy for Children With Thalassemia: Total or Partial Splenectomy, Open or Laparoscopic Splenectomy

Al-Salem AH. *J Pediatr Hematol Oncol.* 2016 Jan;38(1):1-4.

doi: 10.1097/MPH.000000000000121.

The laparoscopic partial splenectomy in pediatric patients: the role of pre-operative imaging and technical details

Prof. Mario Lima

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Figure 1 VR rendering of a splenic cyst. After 3D reconstruction of the different structures (a), it's possible to focus on vascularization (b).

The surgical anatomy is very important in splenic surgery: the major blood supply to the spleen comes from the splenic artery, which originates from the celiac trunk. It splits up into a superior and an inferior body. A single splenic artery is present only in 0.8% of patients, while 86% have, two polar splenic arteries with four segmental ones, that in partial splenectomy may be very useful. We must also focus our attention on the means of fixation of the spleen, such as: the gastro-splenic, pancreatic-splenic and spleno-colic ligament. Do not forget the spleno-phrenic ligament that sometimes in total splenectomy makes the procedure more difficult. Preoperative imaging needs to be carefully evaluated to obtain information regarding the diagnosis, the size of the spleen, its anatomical relationships as well as its vasculature. Ultrasound can help in determining the size, the type of lesion but not the vasculature. Magnetic resonance imaging (MRI) can verify the contents of a cystic lesion and show us the parenchyma and its relationship with surrounding structures.

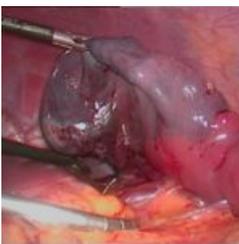


Figure 2 After ligation of the feeding branches, the portion that has to be removed becomes ischemic

In patients with a specific indication to partial splenectomy, CT scan with the angiographic study constitutes a useful tool for the assessment of the vascularization. Both arterial supply and venous drainage have to be taken into account in preoperative imaging, especially when a partial splenectomy is required. In the last years a new technology has been developed among the preoperative imaging: the virtual reconstruction (VR rendering). This technology allows to obtain a three-dimensional high-contrast, enhanced vascular structure study with the possibility to exclude nearby organs, focusing on the anatomic reconstruction of the spleen and its vessels. In patients with a specific indication to partial splenectomy, it's always appropriate to inform the parents of the possibility to proceed to a total splenectomy, especially in case of massive bleeding which can be difficult to control. In partial splenectomy we preserve more or less the 25% of normal splenic parenchyma. In the past years it was only described to treat Gaucher disease, nowadays it is most often considered, also in hereditary spherocytosis. The knowledge of vascularization allows to perform precise ligatures in order to obtain optimal demarcation of the splenic tissue that has to be removed (Fig. 2). Both the upper and the lower pole may present anatomical variants of the blood supply not originated from the hilum. In particular the upper pole may receive its blood supply by the short gastric vessels, while the lower pole, from the ascending branch of the left gastro-epiploic artery. These vascular branches are useful in the subtotal splenectomy, where 85-90% of the splenic tissue is removed. Do not forget also the phreno-splenic and colo-splenic ligaments that can keep in place the spleen. In unroofing technique, the cyst is emptied and the periferic wall is removed. Unfortunately, the major problem is the inner layer, behind which there may be other small cysts, which once decompressed, can develop. The persistence of a part of the cyst appears to favor recurrences (43-64%). One of the ways to prevent recurrences is the removal of the remaining inner layer, but it is technically more difficult. The patient should be placed on anti-Trendelenburg and semi-lateral decubitus position. The trocars are placed along an imaginary semicircular line: a balloon anchoring 10/mm trocar is inserted in the umbilicus by an open approach and pneumoperitoneum is established with insufflation of 0,5- 11 / min flow at an eight -twelve mm / Hg pressure . I prefer to use a 10 mm direct optic at zero degrees; two 5 mm operating trocars

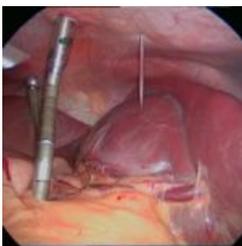


Figure 3 A cannula can be used to empty the cyst and facilitate the removal

are inserted respectively below the right and the left costal margin, medially and laterally to the midclavicular line; a fourth 3-5 mm trocar is inserted under the xiphoid process, to retract and to allow the best exposure of the spleen. When we plan a partial splenectomy, such as in case of cystic lesion as showed in Figure 3, it is very useful to empty the cyst with a cannula, a Deflux and/ or bone marrow aspirate needle.

Sometimes aspiration can be impaired by the presence of septa within the cyst so it is possible to use 5 mm laparoscopic needle that perform this task. Technically we have to check, identify and dissect the ligaments, according to the site of the lesion and consequently dissect the gastro-epiploic branches and eventually the short gastric vessels. Thus, in the case of a lower

pole lesion we must proceed with the section of the spleno-pancreatic ligament. Nowadays, with new devices, it is much easier: among the available instruments for parenchyma and vessel sealing, I prefer the one that uses a combination of pressure, supplied from the hand piece and radio frequency (RF) applied to target tissues. This instrument can have two types of tips, the Dolphin tip and the Maryland tip, in my opinion the Maryland is much more precise: it can also be used as a dissector and nontraumatic forceps. We can apply these devices on vessels up to 7 mm: it is very difficult to measure the real size of the vessels, thus the surgeon experience is significant. Furthermore, we have always to be ready to a different hemostasis technique such as clips and ligatures. After the anterior surface of the splenic hilum is exposed, I prefer not to use ultrasonic technology because the branches remain hot for a long time and this can be dangerous to the surrounding organs such as the pancreas, spleen etc.

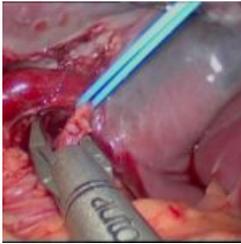


Figure 4 Once isolated the vessel, a rubber band is put around allowing to better handle it

Ligatures, clips, stapler, or if you feel comfortable, the Ligasure, can all be used to manage the main vessels. The control of the vascular supply is the most delicate step of the procedure. After isolation of the vessels, I use to put a rubber band around each vessel and suspend them: this allows a better visualization and avoids injuries to unrecognized posterior vessels (Fig. 4). Once the vessels are isolated and ligated, the related parenchyma becomes ischemic: the surgeon can identify the transection line, 1 cm from the demarcation line on the devascularized side of the spleen (Fig. 5).

Partial splenectomy is necessary in splenic cyst because there could be those smaller in contiguity with the greater one that can lead to recurrences. In these cases we still use the hemostatic radio-frequency technology because it allows a quite bloodless section. Others strategies are available to ensure complete hemostasis of the section surface such as fibrin glue. The exteriorization of the operative specimen must be done in a protected manner by insertion of an endobag followed by the extraction from the umbilicus. The Pfannenstiel incision is rarely necessary to exteriorize the specimen after a partial resection; it is also possible to use a Morcellator for larger lesions, although it is dangerous because of the risk of abdominal splenosis. In our experience, bleeding has been the first cause of conversion. The preparation and the section of ligaments is very important and should be aimed at preservation of the ligaments closest to the splenic portion which must not be detached: this surgical step can be performed faster thanks to the use of modern devices. The preparation and section of the vessels must be meticulous, always separating the afferent artery from the vein (pre-operative work-up).

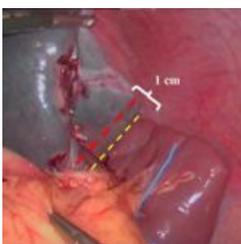


Figure 5 The section line (red line) has to be at about 1 cm from the demarcation between ischemic and non ischemic parenchyma (yellow line)

The section of the parenchyma is made simpler by the radiofrequency device, but it remains a technically difficult procedure because of possible bleeding from unrecognized vessels; It's mandatory to be cautious and prepared to handle any bleeding complications.

Partial splenic surgery: literature data

Prof. Girolamo Mattioli

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It's very difficult to discuss literature data on partial splenic surgery because there are no randomized trials and /or controlled prospective, double-blind studies, and I think that we can not take a definite position on partial splenectomy. Thus, we have many heterogeneous data and it is clear that we must never take out a healthy spleen. Founded on our experience partial splenectomy does not see our approval in hematological disease. As Prof lima said before, in the patient with a splenic cyst we prefer to perform un-roofing and omentopexy within the cyst. The hemato—oncologists want a splenectomy for purpura and spherocytosis: in these cases there are data in the literature for the partial splenectomy, but are not randomized. In the simple cyst treatment , we have the interventional radiologists who can make an endovascular ischemia; fenestration may be an alternative, if the radiologists cannot treat it. Our hematologists do not require a partial splenectomy because they are much more worried about the possible persistence of the disease, compared to the risk of sepsis, that still exists and they know that there is a high-risk. In the literature of the last five years, I realized that there are merely opinions of experts.

Before planning a partial resection, it is very important to do a vascular preoperative planning; the aim is to preserve the spleen, but the risk of cauterization of the capsular surface exposes to a possible adhesions syndrome. When we talk about the outcome of partial resections, we must discuss the long-term results, before concluding that a procedure is better than another, especially in immuno-compromised patients. Another aspect, as it has already said Professor Lima, is the random vascularization of the spleen, where we have internal anastomoses between the upper pole and the center that allow the post-trauma tissue regeneration. When we have a splenic rupture near the hilum, it undergoes healing, perhaps with outcome in a pseudo cyst, which can have other smaller cysts, but fibrous, not mucosal; then we prefer the un-roofing. I have much experience in the hemi-nephrectomies with partial resection, where you can manage it, because we have a clear vascularization; attention must be paid to the treatment of the vein that is very gentle compared to arteries. In these cases, I prefer to use the laces for tying the vessels. Another aspect is that with the radio frequency technology, I cannot resect the parenchyma because the new Maryland is active for the release of energy and therefore for sealing tissue, only when it has a closed-bite; on the other hand this technology is not born for the tissues, but for the vessels. So in the case of a hemi-splenectomy, I would be afraid to use this device. This movie shows you that after vascular ligatures the upper pole has become ischemic only thanks to the section of the short gastric vessels; the lower pole has not become ischemic with the ligature of the lower pole vessels. So for an upper polar hemi-splenectomy, you just need to dissect the short gastric vessels; to obtain an ischemia of the lower pole it is not adequate the ligature of the inferior vascular pedicle. In conclusion, we may adfirm that we don't advise hemi-splenectomy for hemato—oncological diseases; partial resection for superior polar cysts perhaps and if they are in the lower pole, it is very difficult not to sacrifice the hilum. I conclude: literature does not give strong messages. The experience is crucial to try to preserve the splenic tissue.

Recommended lectures

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Sonography of the spleen.

J Ultrasound Med. 2011 Sep;30(9):1281-93.

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3D virtual rendering before laparoscopic partial splenectomy in children.

J Pediatr Surg. 2013 Aug;48(8):1784-8.

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Partial versus total splenectomy in children with hereditary spherocytosis.

Surgery. 2013 Oct;154(4):849-53; discussion 853-5.

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Single-incision pediatric endosurgery (SIPES) splenectomy: a case-control series.

J Laparo endosc Adv Surg Tech A. 2013 Feb;23(2):162-5.

Rice HE, Crary SE, Langer JC, Kemper AR; Splenectomy in Congenital Hemolytic Anemia Consortium.

Comparative effectiveness of different types of splenectomy for children with congenital haemolytic anemias.

J Pediatr. 2012 Apr;160(4):684-689.e13.

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Anterior versus postero lateral approach for total laparoscopic splenectomy: a comparative study.

Int J Med Sci. 2013;10(3):222-9.

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Laparoscopic cystectomy for splenic hydatid cyst: a case report.

J Pediatr Surg. 2011 Nov;46(11):e1-3.

Buesing KL, Tracy ET, Kiernan C, Pastor AC, Cassidy LD, Scott JP, Ware RE, Davidoff AM, Rescorla FJ, Langer JC, Rice HE, Oldham KT.

Partial splenectomy for hereditary spherocytosis: a multi-institutional review.

J Pediatr Surg. 2011 Jan;46(1):178-83.

Vasilescu C, Stanciulea O, Tudor S.

Laparoscopic versus robotic subtotal splenectomy in hereditary spherocytosis. Potential advantages and limits of an expensive approach.

Surg Endosc. 2012 Oct;26(10):2802-9.

Vasilescu C, Stanciulea O, Arion C.

Laparoscopic subtotal splenectomy in hereditary spherocytosis.

Surg Endosc. 2007 Sep;21(9):1678.

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Laparoscopic subtotal splenectomy in hereditary spherocytosis : to preserve the upper or the lower pole of the spleen?

Surg Endosc. 2006 May;20(5):748-52.

Imuro Y, Okada T, Sueoka H, Hai S, Kondo Y, Suzumura K, Fujimoto J.

Laparoscopic management of giant splenic true cyst with partial splenectomy: a case report.

Asian J Endosc Surg. 2013 Aug;6(3):226-30.

Schier F, Waag KL, Ure B.

Laparoscopic unroofing of splenic cysts results in a high rate of recurrences.

J Pediatr Surg. 2007 Nov;42(11):1860-3.

Podevin G, Victor A, De Napoli S, Heloury Y, Leclair MD.

Laparoscopic splenectomy: comparison between anterior and lateral approaches.

J Laparoendosc Adv Surg Tech A. 2011 Nov;21(9):865-8.

Wang X, Wang M, Zhang H, Peng B.

Laparoscopic partial splenectomy is safe and effective in patients with focal benign splenic lesion.

Surg Endosc. 2014 Dec;28(12):3273-8.

de la Villeon B1, Zarzavadjian Le Bian A, Vuarnesson H, MunozBongrand N, Halimi B, Sarfati E, Cattan P, Chirica M.

Laparoscopic partial splenectomy: a technical tip.

Surg Endosc. 2015 Jan;29(1):94-9.

DISCUSSION

Prof. Papparella: At present what are the indications or the future prospects for the partial splenectomy? There is actually a benefit for the immunological function and prophylaxis in these patients or there is still no agreement with this argument?

Prof. Ruggiero: The partial splenectomy preserves immune function, so theoretically these patients do not require vaccination and antibiotic prophylaxis

Prof. Mattioli: The literature does not have large series and 30% of the patients return to the operating room for a total splenectomy; the surgical risk of relapse is very high. In hematological diseases at present there are no studies in the literature that present an absolute indication for partial splenectomy. Our hematologists are much more concerned about the persistence of the disease.

Prof. Cozzi: We do not have a great experience in laparoscopy, but in open surgery and even in the partial we vaccinate them as well as total splenectomy. Our hematologists have reviewed the indications and I think that one of these could be an age under five years old, and basically it becomes a bridge therapy: it is true that these patients have a risk to undergo a total splenectomy, but they do later in life. Concerning on literature, I remember to the audience

that there is the experience of the Toronto group, which is very wide and then I think there is an hematological trend in indications of the partial splenectomy. I agree with Professor Lima on splenic cysts because these have a high recurrence rate, and we must try to schedule a surgery that must be radical. The rendering techniques and the pre-operative study can help us a great deal. I think that the partial splenectomy should be considered in the treatment of cysts, but always has to be radical.

Prof. Lima: Patients who undergo surgery of the spleen should be vaccinated, since it is not known in these patients as surgery, even in the course of partial splenectomy, is definitive ; in these cases we do not have the clear limits for vascular anastomosis; I remember also that in the TAC evaluation, we must also look at the parenchyma, for other small cysts; if you have difficulty, don't be afraid to convert: is better to convert the procedure rather than enter into a vortex of complications

Dr. Porreca: I report an anecdote from Dr. Holcomb, who told us that a child , where a partial splenectomy was performed, died from a ruptured spleen as a result of trauma. I think that the simple cysts are a good indication of this type of surgery; I have observed a case of disappearance after drainage and alcoholization but in the literature there are also cases of sepsis after Interventional radiology.