First experience with single incision laparoscopic surgery in Slovakia: Concomitant cholecystectomy and splenectomy in an 11-year-old girl with hereditary spherocytosis

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Aims. Hereditary spherocytosis is an autosomal dominant inheritance disorder of the red blood cell membrane characterized by the presence of spherical-shaped erythrocytes (spherocytes) in the peripheral blood. The main clinical features include haemolytic anemia, variable jaundice, splenomegaly and cholelithiasis caused by hyperbilirubinemia from erythrocyte hemolysis. Splenectomy does not solve the congenital genetic defect but it stops pathological hemolysis in the enlarged spleen. If gallstones are present, it is appropriate to perform cholecystectomy at the time of splenectomy, although the patient has symptoms of gall bladder disease. We present the case of single incision laparoscopic surgical (SILS) concomitant splenectomy and cholecystectomy performed with conventional laparoscopic instruments in an 11-year-old girl with the diagnosis of hereditary spherocytosis.

Methods. A 2 - 3 cm umbilical incision was used for the placement of two 5 mm trocars and one 10 mm flexible videooscope. Conventional laparoscopic dissector, grasper, Ligasure, Harmonic Ace and hemoclips were the main tools during surgical procedure. We prefer Single Incision Laparoscopic Surgery Foam Port (Covidien) as the single umbilical device for introduction into the abdominal cavity. First, we performed cholecystectomy, then the gallbladder was put aside over the liver and after that we performed splenectomy. To remove the detached spleen and gallbladder, a nylon extraction bag is introduced through one of the port sites. The spleen is than morcellated in the bag with forceps and removed in fragments. After that we removed them and the umbilical fascial incision was closed.

Result. Splenectomy is the only effective therapy for this disorder and often it is performed in combination with cholecystectomy. Conventional surgery requires a wide upper abdominal incision for correct exposure of the gallbladder and spleen. Our experience shows that SILS splenectomy and cholecystectomy is feasible even in young children and despite the small number of cases in the world, we consider the combined laparoscopic approach safe and effective for the treatment of hereditary spherocytosis.

Conclusion. According to actually published guidelines, the laparoscopic approach to concomitant splenectomy and cholecystectomy is recommended, but it depends on the availability of appropriately trained surgeons and suitable equipment.

Key words: single incision laparoscopic surgery, hereditary spherocytosis, splenectomy, cholecystectomy

INTRODUCTION

Hereditary spherocytosis (HS) refers to a group of heterogeneous inherited anemia that is characterized by the presence of spherical-shaped erythrocytes called spherocytes, on the peripheral blood smear. The abnormal morphology and shorter lifespan of the erythrocytes in HS are attributable to a deficiency or dysfunction in one or more of the proteins that compose the red blood cell membrane cytoskeleton whose role is to maintain the shape, deformability and elasticity of the erythrocyte1,2. Hereditary spherocytosis is the most common congenital hemolytic anemia in Caucasians, with an estimated prevalence ranging from 1:2000 to 1:5000. Approximately 75% of cases display an autosomal dominant pattern of inheritance, the remaining comprise recessive forms and de novo mutations3. The main clinical features of hereditary spherocytosis are haemolytic anaemia, variable jaundice, splenomegaly and premature cholelithiasis. Although the diagnosis of HS is often made in childhood and young adulthood, it may be diagnosed at any time of life including old age4,5.

The two most often affected organs by HS are the spleen and gallbladder. With regard to its structure and function, the spleen is considered as two separate organs. The white pulp of the spleen is an important part of the
immune system, whereas the red pulp is connected to the bloodstream and is engaged in phagocytosis of senescent, damaged, or genetically altered (e.g. hereditary spherocytosis) red blood cells. The extraordinarily splenic blood microcirculation is basic for red pulp’s very sensitive blood purge. Some of arterioles and arterial capillaries end blindly while the blood freely flows out into the three-dimensional network of connective tissue splenic cords of Billroth, with numerous macrophages. The blood elements with undisturbed elasticity of cell membrane subsequently return to the blood circulation by penetrating through the small openings in the walls of specialized capillaries (splenic venous sinuses) (ref.\(^7\)-\(^10\)). The altered erythrocytes in hereditary spherocytosis are vulnerable to splenic sequestration and subsequent lysis and cause the characteristics of this disease such as jaundice, anemia, splenomegaly, and cholelithiasis. According to actually published guidelines, the laparoscopic approach to splenectomy is recommended, but this is dependent on the availability of appropriately trained surgeons, and suitable equipment. New guidelines also recommend, when splenectomy is indicated in children, ideally it should be done after the age of six\(^11\).

In HS is a high incidence of pigmented gallstones among patients with spherocytosis, similar to other hemolytic anemias. Cholelithiasis is caused by hyperbilirubinemia from erythrocyte hemolysis and affects 50% of unsplenectomized patients who have hereditary spherocytosis. Splenectomy alone is sufficient treatment in the absence of gallstones. If gallstones are present, it is appropriate to perform cholecystectomy at the time of splenectomy, although if the patient has symptoms of gall bladder disease, most surgeons will remove the gall bladder at the same time as the splenectomy\(^6\),\(^12\). According to Guidelines for the diagnosis and management of hereditary spherocytosis – 2011 update, when stones are incidental findings without symptoms, the value of cholecystectomy remains controversial\(^11\). As per our experience, single incision laparoscopic surgery (SILS) can be used effectively to perform simultaneous splenectomy and cholecystectomy in patients with hereditary spherocytosis\(^13\).

In this article we present our experience with an 11-year-old girl who underwent combined SILS splenectomy and cholecystectomy for hereditary spherocytosis with positive family history, splenomegaly and cholelithiasis.

**Patient**

The preoperative diagnosis and indications for splenectomy in our patient, an 11-year-old girl, were established by a pediatric hematologist. The patient received preoperative vaccination with polyvalent pneumococcal, meningococcal and Haemophilus influenza vaccines. In this case gallstones were present on ultrasonography, so it was appropriate to perform cholecystectomy at the time of splenectomy which was indicated at the Department of Pediatric Surgery of the Children’s Medical Hospital in Bratislava, Slovakia.
Instruments

The advent of newer laparoscopic trocars and instruments allow us to perform scarless surgery. All instruments enter the abdominal cavity through a single incision (Fig. 1). There is just one incision within the belly button, which leads to less pain and improved cosmetic outcome. Available instruments can be articulating, curve-shaped, or conventional rigid laparoscopic devices. Instruments which are roticulating and provide several degrees of freedom help us to overcome the main problem - triangulation reduction tools and allow us to perform these operations rather than the standard 3 or 4 incision laparoscopy. In this case, the SILS splenectomy and cholecystectomy was performed with conventional laparoscopic instruments. A 2 - 3 cm umbilical incision was used for the placement of two (5 mm) trocars and one (10-mm) flexible videoscope (Fig. 2). Conventional laparoscopic dissector, grasper, Ligasure and Harmonic Ace were the main tools during our surgical procedure.

Technique of creation of pneumoperitoneum

SILS concomitant splenectomy and cholecystectomy was performed by two attending surgeons. Patient underwent general endotracheal anesthesia. The patient was placed in a supine - frog position. The main surgeon usually stands between the patient’s legs, which offer a more ergonomical approach. The basic surgical steps for a SILS are similar to the standard laparoscopic approach. The essential differences are in the way we enter the abdominal cavity, the location of SILS port and laparoscopic instruments, which are used. The umbilicus is the anatomic area for access to the abdominal cavity from both a practical and cosmetic point of view. The skin incision on the lower border of the navel about 2/3 of its circumference then the umbilicus is mobilized and completely everted as in umbilical hernia repair. The peritoneal cavity is opened through the vertical incision of the linea alba using the scissors incised vertically along its midline. Hemostasis should be precise to prevent postoperative hematoma. The fascial incision is extended up and down to create a 2 to 3 cm defect in the linea alba. On the margin of the mini-laparotomy, we place non-absorbable stitches. This allows traction of the abdominal entire during manual SILS port insertion into the abdominal cavity. This incision (mini-laparotomy) later proves to be ideal for relatively large specimen extraction. We prefer Single Incision Laparoscopic Surgery Foam Port (Covidien) as the single umbilical device for introduction of the instruments into the abdominal cavity (Fig. 1). The Covidien SILS port is pushed via the incision starting with the upper more rigid edge (where is the insufflations tube located) and working to the lower softer edge. A flexible videoscope (diameter 10 mm; 0 degrees) is introduced to ensure port position in the abdominal cavity. Pneumoperitoneum was established to 12-14 mm Hg and an adequate workspace has been created. Two additional low-profile 5-mm ports are then placed. It becomes evident that conflicts exist between the hands and instruments of surgeon and assistant. Also the surgeon needs to use his left hand though

Fig. 4. Dissection of the distal polar vessels with Harmonic scalpel.

Fig. 5. If the artery is first divided, spleen size and tissue turgor may decrease in important ways and make splenectomy easier.

Fig. 6. Finally removing of both organs with using an endo-bag.
ordinarily he would prefer the right one to clips or to cut as a classic laparoscopy.

**SILS cholecystectomy**

The assistant’s Grasper is used to elevate the fundus of gallbladder over the liver to ensure optimal exposure of the triangle of Calot. The surgeon retracts the infundibulum of the gallbladder laterally to create appropriate orientation of the cystic and common ducts. Once the anatomy is visualized, cystic duct is doubly clipped on his proximal and distal side and than divided (with Metzenbaum scissors) in the middle of the endoscopic clips (Fig. 3). The cystic artery and cystic duct are clipped separately with a standard 5-mm clip applied. The cystic artery can be as well interrupted by Covidien Ligasure vessel sealing device (Valleylab). Dissection of gallbladder from his liver bed is performed as a normal retrograde cholecystectomy using a monopolar Hook or Harmonic Ace (Ethicon surgery). Hemostasis is controlled with the cautery. Than the gallbladder is put aside over the liver.

**SILS total splenectomy**

Turning the patient to the right (semidecubitus position with the left side elevated was achieved by tilting the table during the procedure), so that the spleen fell in to the surgical field. Dissection begins with mobilization of the splenic flexure of the colon and division of the splenocolic ligament and distal polar spleen vessels using the Harmonic Ace (Fig. 4). This allows it to be retracted cephalad. Currently available vessel division tools for the control of the short gastric vessels include the ultrasonic dissector, hemoclips and Ligasure device. After the short gastric vessels are divided, the splenic pedicle may be carefully dissected free superolaterally and anteroposteriorly. For larger hilar vessels (Fig. 5), we applied the Ligasure at 2 points adjacent to each other, and divided only on the seal nearer the spleen. (Contrary to the popular belief, it is not useful to apply energy with the Ligasure at the same site more than once. Moreover, one can risk charring and tearing the vessel with this technique). An endostapler can also be used to divide the vessels in the hilum. The patient had avulsion of a hilar venous branch. An episode of bleeding at the splenic hilum was controlled via SILS. This vessel was controlled with a 5-mm clip applier, and conversion was not necessary. The patient received 1 U packed red blood cell transfusion. There were no other complications.

After the blood vessels in hilum have been ligated and divided, the completely devascularized spleen was pendent on a small cuff of avascular superior pole splenophrenic ligament, which was then divided.

**Finally removing both organs**

To remove the detached organs, a nylon extraction bag was introduced through one of the port sites. The bag was opened within the abdominal cavity and the specimen was introduced into the endo-bag, after internal fragmentation, the bag was closed and exteriorized through the umbilical incision - mini-laparotomy. It is very easy to
remove the specimen in the endo-bag through the SILS umbilical incision (Fig. 6).

After we removed the gallbladder and the spleen, the umbilical fascial incision was closed. We prefer to close the fascia with interrupted non-absorbable sutures, then the skin is closed with interrupted sutures. The drain was placed through the mini-laparotomy (Fig. 7), the Betadine® unguent and sterile dressing was applied on the umbilicus and the patient was transported to the recovery room.

The main benefit of an SILS approach to splenectomy and cholecystectomy appears to be cosmetic because no visible scar results, especial in a case of young girl (compare Fig. 8 and Fig. 9).

Histological evaluation of removed organs

The fragments of the spleen and gallbladder were after formalin-fixation and paraffin-embedding examined by a pathologist and histologist. In classical histological staining with hematoxylin and eosin, the white pulp of the spleen had a normal appearance, but the splenic red pulp had marked blood congestion in the splenic (Billroth’s) cords and we also observed hyperemic blood sinuses (Fig. 10).

The gallbladder mucosal folds were atrophic and the covering epithelium was damaged. The mucosa contained numerous diverticula called Rokitansky-Aschoff sinuses, which may be associated with chronic cholecystitis and cholelithiasis. We observed numerous lymphoid follicles as other morphological signs of chronic cholecystitis and cholelithiasis (Fig. 11).

DISCUSSION

More than 4,500 reports of SILS procedures have been published14 and we may see the same phenomenon occur with the application of single-site laparoscopy. SILS technique was established to facilitate larger specimen retrieval without the need of additional fascial and skin incisions. If multiple procedures are performed at the same time, patients also undergo a solitary hospital admission, preoperative evaluation and anesthesia exposure. Single-incision surgery has been applied to a number of procedures in adults such as appendectomy, cholecystectomy, gastrectomy, adrenalectomy, colorectal procedure, etc.15-19.

We have experienced positive results from performing laparoscopic splenectomy and cholecystectomy at one time for treating hereditary spherocytosis that was combined with cholelithiasis. The operation procedure can be difficult to achieve because all instruments come from one direction19. Our operation was a rare SILS procedure, when spleen is removed concomitant with the gallbladder (Table 1).

Using instruments of different lengths as well as angled scope prevent clashing between tools and hands of the surgeon and assistant. The use of flexible scope can reduce the possibility of having the camera in-line with the instruments but requires a skilled assistant to operate the telescope. The operative time depends not only on surgeon experience but also on anatomic conditions, the method of blood vessel closing, manner of spleen removal, the size of the spleen and other factors21-23. In spite of all these reasons, for experienced laparoscopic surgeons SILS is now easier and there is evidence with appropriate training in rapid decrease in operative times as surgeons acquire experience with the technique. Then overall operative time decreases to a level comparable with standard laparoscopic surgery24,25.

Potential postoperative complications with SILS splenectomy and cholecystectomy include wound seroma or ventral hernia. Mobilization of umbilical stem creates a place for eventual fluid collection and infection26. This
has been reported in several cases of pediatric SILS appendectomy for complicated appendicitis but not in cholecystectomy or splenectomy.27,28

Guidelines on hereditary spherocytosis published in 2011 reflect current opinion on the surgical management in children with hereditary spherocytosis. Further potential long term hazards of splenectomy were recognized. So there are recommendations for partial splenectomy. It may be beneficial but needs further follow-up studies.11

Classically, laparoscopic splenectomy is performed using 4 ports: 10 or 12 mm. Cholecystectomy is performed before splenectomy with or without the additional 1 or 2 ports in right upper quadrant. The main benefit of an SILS approach to splenectomy and cholecystectomy appears to be cosmetic because no visible scar results.14

CONCLUSIONS

Splenectomy is the only effective therapy for hereditary spherocytosis and often it is performed in combination with cholecystectomy in adults. Conventional surgery requires a wide upper abdominal incision for correct exposure of the gallbladder and spleen. Laparoscopic cholecystectomy and splenectomy have been performed safely worldwide. We report our experience with a child who underwent combined SILS splenectomy and cholecystectomy for hereditary spherocytosis.

Our report shows that single-incision splenectomy and cholecystectomy is feasible even in young children and despite the small number of cases in the world, we consider the combined laparoscopic approach safe and effective for the treatment of hereditary spherocytosis.

Laparoscopic splenectomy has become a gold standard in the treatment of spleen disorders related to hematologic diseases. Increasing laparoscopic surgery experience and improved new vessel sealing equipment have led to a decrease in number of ports in laparoscopic surgery and to operations from one incision.

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