

## Concomitant laparoscopic cholecystectomy and laproscopic assisted splenectomy for surgical management of hereditary spherocytosis

Khairalleh Muzhir and Hussein Ali Albudairy

Medical College /Wasit University

### عملية رفع المرارة والطحال بالناظور لمريضة مصابة بتشوة خلقي في كريات الدم الحمراء

خير الله مزهر و حسين علي البديري

كلية الطب- جامعة واسط

#### المستخلص

الجراحة المنظارية من الجراحات المتقدمة خصوصا عمليات رفع المرارة والطحال. اجريت عملية رفع الطحال والمرارة بالناظور لامرأة في سن الخامسة والعشرين تعاني من تضخم الطحال مع حصاة المرارة مع فقر دم شديد لان المريضة تعاني تكسر الكريات الحمراء بسبب تشوه الكريات الحمراء بالاضافة ان المريضة بالاشهر الاولى من الحمل اجريت العملية وتكللت بالنجاح المريضة والطفل بصحة جيدة

#### Abstract

Laparoscopic splenectomy is rapidly becoming a common treatment modality in the surgical management of hematological processes involving the spleen .Hereditary spherocytosis is the most common red blood cell membrane disorder and its diagnosis is often associated with haemolytic crisis and premature cholelithiasis .our patient 25 year old presented with cholelithiasis ,splenomegaly and anemia and because of persistent anemia and gastrointestinal symptoms the patient underwent laparoscopic cholecystectomy and splenectomy in pregnant women (1<sup>st</sup> trimester)

#### Introduction

Hereditary spherocytosis is a disorder of the red blood cell membrane protein that leads to sphere shaped red blood cells which are less resistant to stress and rupture easily leading to chronic hemolytic anemia. The disease may be mild need no treatment or moderate disease treated with folic acid 5 mg/day supportive treatment and regular annual checkup or severe disease warrants frequent hematological supervision and splenectomy (1).

#### Laparoscopic splenectomy

##### Contraindication

- cancer,
- large hilar lymph node,
- portal hypertension

- Absolute contraindication massive splenomegaly (craniocaudad length >16 or wt >600 g supermassive >22cm or wt >1600g (2)
- Patient Selection and Pre-operative Planning
- The laparoscopic approach should be considered as a therapeutic option for all patients undergoing elective splenectomy. A few important contraindications to the laparoscopic approach are patients with liver failure with portal hypertension, ascities or unmanageable coagulopathy. In addition, while laparoscopic management of splenic trauma has been reported in the literature, it is not standard of care, and should not be considered in a patient with hemodynamic instability.(3)
- Splenic size is a key factor in determining the feasibility of the laparoscopic approach, and should be determined prior to taking the patient to the OR. Splenic size has been reported in terms of length of greatest dimension in centimeters or in terms of weight. There is no consensus on how to categorize varying degrees of splenomegaly (4) Generally, normal spleens are <12 cm, moderately enlarged spleens are 12-20 cm and severely enlarged spleens are >20 cm. Special consideration should be given to spleens >20 cm, which are considered to be “giant” sized spleens, and are really difficult to remove laparoscopically due in part to limitations in instrumentation and visualization. More importantly, there is the difficulty to, at first, establish a working space (pneumoperitonium) in an abdominal cavity already stretched with a massive spleen and, secondly, trying to insert a massive spleen into an adequately sized extraction bag in such a restricted working environment. Of course, the greater the size, the greater the difficulty of the case, and more technical ability is required to safely remove the spleen laparoscopically.(5)
- In addition to splenic size, imaging should also be reviewed for the presence of accessory spleens. Some have suggested that a major pitfall to laparoscopic splenectomy is the limited ability to explore the abdomen for accessory spleens (6). Given that the majority (>80%) of accessory spleens are in the hilum near the vascular pedicle or omentum near the spleen this concern has not been shown to be clinically significant. Nevertheless, as the amelioration of certain disease processes is dependent on the removal of all splenic tissue, the review of preoperative imaging is prudent. Some have advocated obtaining liver-spleen nuclear medicine scintigraphy for this purpose, but it is likely an unnecessary adjunct to routine CT scanning.(7)
- In the elective setting, patients should be counseled regarding risks of splenectomy, including the risk of infection from encapsulated organisms. Appropriate vaccination against Pneumococcus and H. influenza should be administered weeks prior to surgery. Meningococcal vaccination should be offered to pediatric patients or younger patients who live in dormitories (college, military).(8)
- Patients with thrombocytopenia should generally be maintained with a platelet number above 30,000, although internal bleeding is rare unless there are persistent counts <5000. In the case of ITP, transfusion of platelets should be avoided until after surgery or during surgery after “inflow” vascular isolation (9). If it is necessary to increase platelet count in

these patients prior to surgery, usually a steroids bolus or parenteral IVIG can boost platelet count prior to surgery. Similarly, ideally hemoglobin should be at least 10 g/dl prior to surgery.(10)

- As these patients may be chronically dependent on steroids, adrenal insufficiency may be a concern and peri-operative stress dose steroids may be considered.(11)

### Advantages of laparoscopic splenectomy

Laparoscopic splenectomy is widely used as the preferred approach because it is less traumatic, engenders less postoperative pain, and is associated with fewer wound infections and other complications leading to shorter hospital stays, more rapid convalescence, and more rapid return to work, all of which contribute to lower costs (12). Cosmesis is improved compared with the open approach. Laparoscopy does not increase the frequency of missing accessory spleens. Consequently, laparoscopic splenectomy has been embraced as the “gold standard” but should only be performed by experienced operators. The conversion rate to open splenectomy , ranges from 5% - 15% (13) .

### Case report

A 25 years old pregnant women (1<sup>st</sup> trimester) presented with recurrent abdominal pain at the right hypochondrium and repeated vomiting and history of severe pallor and recurrent yellowish discoloration of the eyes and generalize weakness .Examination revealed a young lady with pallor and jaundice and with massive splenomegaly.

Laboratory investigation showed

Hb 5.0 gm/dl

WBC 6.700 cell/mm<sup>3</sup>

Neutrophil = 46% Lymphocyte= 48% Monocyte =4% Eosinophil =2%

Platelet = 240.000 /mm<sup>3</sup>

RBC hypochromic microcytic with anisocytosis and poikilocytosis(tear drop ,target cell,pencil.shaped) poikilocytes.

Normal Platelet

WBC lymphocytosis. No immature cells seen

ESR 5 mm/hour

Blood glucose 95.5 mg/dl

Blood urea 23.4 mg/dl

S.Creatinine <0.5 mg/dl

TSB 4.29 mg/dl

Direct serum bilirubin 0.57 mg/dl

Indirect serum bilirubin 3.72 mg/dl

S.GPT 16.4 U/L

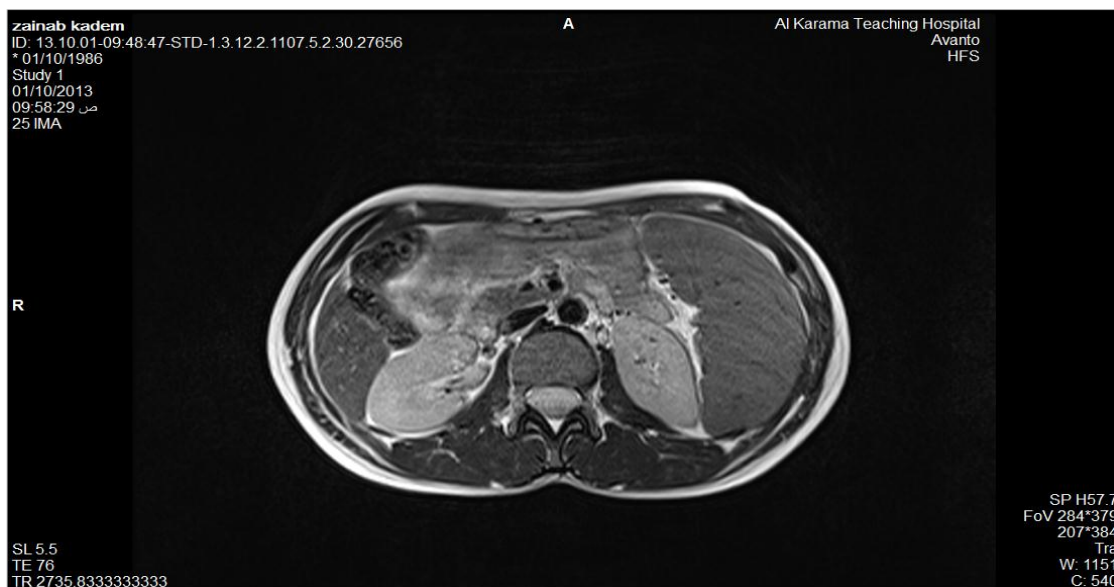
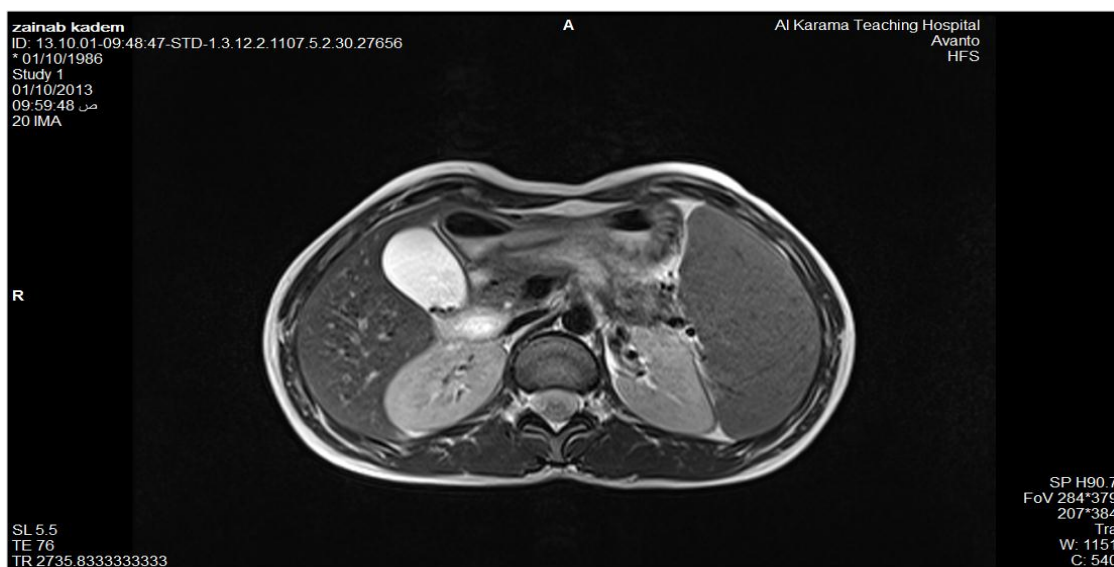
S.GOT 24.9 u/l

S .ALK.Pho 58.4U/L

Blood group and Rh B+

Clotting time 2 minutes

Bleeding time 1.5 minutes



**Figures (1-2): CT scan of abdomen and pelvis show multiple gall stons and huge splenomegaly**

### Preoperative preparation

The preoperative diagnosis and indications for splenectomy were established by a hematologist as a case of spherocytosis and gall stone with massive splenomegaly and when become pregnant developed hemolytic crisis and severe anemia and need multiple blood transfusion a decision to do laparoscopic splenectomy was taken .She was transfused blood to correct anemia. The patient received preoperative vaccination with polyvalent pneumococcal, meningococcal and *Haemophilus influenzae* vaccines after two weeks splenectomy was done after correction Hb and coagulopathy as well as obtaining informed consent and the family agreed with risk of abortion subcutaneous heparin injections was given and compression stockings was put. A Foley catheter and nasogastric tube insertion. Antibiotic prophylaxis was given to decrease post splenectomy sepsis.Under GA in supine position with reverse Trendeling turn to right during splenectomy and left during cholecystectomy anterior approach –it is only used in special circumstances in very large spleen and when other procedures need to be performed cholecystectomy like in our case also performing complete exploration for accessory spleens.

### Post operative care

- 1-Bloods FBC, U&E evening or next morning as per operation note
- 2- Watch platelet count – may need anticoagulants if increases ++
- 3-Antibiotics as per operation note (Usually IV cephazolin 24hours)
- 4-Chest physiotherapy to prevent chest infection may be required
- 5-Discuss medical alert bracelet
- 6- Ensure vaccination plan is clearly documented on discharge letter – and clarify who will follow this through



Figures ( 3-4): Spleen during surgery and after surgery

## Conclusion

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 surgery and to operations by single port surgery(14). Various authors have advised against splenectomy during pregnancy due to post splenectomy complication like sepsis and thrombocytosis and thrombosis and perioperative morbidity to the mother and the fetus but none of the reported cases had such problems splenectomy can be done at any time but 2<sup>nd</sup> trimester is preferable (15).

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