

EXPERIENCE OF CONCOMITANT SPLENECTOMY AND CHOLECYSTECTOMY IN PATIENTS WITH SICKLE CELL DISEASE

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Sickle cell disease (SCD) is one of the commonly inherited hemoglobinopathies in the Eastern Province of Saudi Arabia,^{1,2} which is known to have an increased frequency of cholelithiasis.³ Patients with SCD from the Eastern Province are known to have splenomegaly that persists into an older age group, and in some into adult life.^{4,5} This is known to be associated with complications which necessitate splenectomy.⁶⁻⁹ When performed separately, elective cholecystectomy and splenectomy have been shown to be safe and effective in patients with SCD.⁶⁻¹⁰ Should cholecystectomy be performed concomitantly with splenectomy in patients with SCD?

Patients and Methods

A total of 112 patients had splenectomy for various hematological diseases at Qatif Central Hospital, over a period of 10 years, from 1986 to 1996. The most common indication for splenectomy was SCD. Seventy-four patients (66%) with SCD had splenectomy. Of these, 17 (23%) had concomitant splenectomy and cholecystectomy. The charts of these patients were reviewed for age at operation, sex, indication for splenectomy, whether gallstones were symptomatic or incidental, postoperative complications and outcome. The results were compared with the remaining patients with SCD, who only had splenectomy.

The diagnosis of SCD was made on the basis of a positive sickling test and hemoglobin electrophoresis (Helena Laboratories Super Z Electrophoresis Kit). The histology of the spleen and gallbladder were obtained from the histopathology report. Preoperatively, all patients were hydrated with intravenous fluids at 1.5 times their maintenance rate, starting the night of the operation and continued postoperatively, until resuming full oral intake. Where necessary, blood transfusions were given pre-

operatively to restore their Hb to 10-12 g/dL and their hematocrit to 30%-40%. No exchange blood transfusions were performed. All patients received polyvalent Pneumovax (0.5 mL of PMY-immune 23, Lederle), as well as prophylactic antibiotics.

Results

Of seventy-four patients with SCD who had splenectomy, 17 (23%) had concomitant splenectomy and cholecystectomy. There were nine females and eight males. Their ages ranged from 12-60 years (mean 24.4 years). Their HbS ranged from 67%-92.3% (mean 77%), and their HbF ranged from 6.2%-33% (mean 21.2%). Their demographic data, as well as indications for splenectomy and cholecystectomy, are shown in Table 1. The indications for splenectomy were hypersplenism in seven, recurrent splenic sequestration crisis in eight, and splenic abscess in two patients. In 11 patients gallstones were symptomatic in the form of recurrent right upper quadrant abdominal pain, while in the remaining 6 (35.3%), gallstones were discovered incidentally during routine ultrasound investigation.

Histology of the spleens showed features of congestive splenomegaly in 15 patients with sclerosis and hemosiderosis in some of them, as well as sickling of red blood cells. In the remaining two patients with splenic abscess, there was evidence of ischemic infarction and abscess formation. In both patients with splenic abscess the causative organism was *Salmonella* group D. Histology of the gallbladder showed evidence of chronic cholecystitis in 13 patients, and it was normal in 4 (23.5%).

Postoperatively there was no mortality, but two patients developed complications: hematoma at the splenic site, which was evacuated surgically, and wound infection in one of the patients with splenic abscess. This gives a postoperative morbidity of 11.8%. The remaining 57 patients had splenectomy only. There were 39 males and 16 females. Their ages ranged from 4-32 years (mean 11 years). Postoperatively, there was no mortality, but seven patients developed postoperative complications: three developed acute chest syndrome and there was one case

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each of subphrenic hematoma, adhesive intestinal obstruction, chest infection and wound infection. This gives a postoperative morbidity of 12.3%.

Discussion

SCD, which is due to homozygous inheritance of the hemoglobin S (HbS) variant, is one of the commonly inherited hemoglobinopathies in the Eastern Province of Saudi Arabia.^{1,2} Patients with SCD in the Eastern Province have characteristically high levels of both total and fetal hemoglobin (HbF).¹ The high levels of HbF, as well as the frequently associated α -thalassemia, are responsible for the so-called benign sickle cell anemia.¹¹

One of the main organs to be affected in SCD is the spleen. Patients with SCD usually have an enlarged spleen during the first decade of life, which as a result of repeated attacks of vaso-occlusion and infarction, undergoes progressive atrophy, leading to autosplenectomy. However, splenomegaly sometimes persists even into adult life. This is the case among SCD patients in Saudi Arabia, where splenomegaly is common even in adults.^{1,4,5,6,8} The reason for this is not known, although a correlation with persistent splenomegaly and high levels of HbF has been suggested.¹² Persistence of splenomegaly in these patients makes them liable to develop complications which are associated with morbidity, and in some with mortality. These complications include hypersplenism,⁶ acute life-threatening splenic sequestration crisis⁸ and splenic abscess.⁹ Splenectomy in these patients is safe and beneficial in reducing their transfusion requirements and its attendant risks, eliminating the discomfort from mechanical pressure of the enlarged spleen, avoiding the risks of acute splenic sequestration crisis and managing splenic abscess.⁶

Cholelithiasis is a well-known complication of SCD. The frequency of cholelithiasis in patients with SCD is variable, ranging from 4% to 55%, depending on the age of the patients and the diagnostic criteria adopted.¹³⁻¹⁶ In the Eastern Province the frequency of cholelithiasis in patients with SCD was 19.3%,³ and like others we found a definite correlation between the frequency of gallstones and increasing age.¹⁴⁻¹⁶ Gallstones in these patients may be symptomatic or asymptomatic, discovered accidentally during routine abdominal ultrasound evaluation. Previously, as a result of a higher incidence of postoperative complications, cholecystectomy in asymptomatic patients with SCD and gallstones was not advisable,^{17,18} but recently, as a result of a better understanding of the pathophysiology of SCD and improved perioperative care, including better anesthetic techniques, surgery became safe, with no mortality and minimal morbidity.^{19,20} Cholecystectomy is the treatment of choice in these patients, even if gallstones are asymptomatic,

TABLE 1. Summary of 17 patients with sickle cell disease who had concomitant splenectomy and cholecystectomy.

Age/sex	HbS	HbF	Indication for splenectomy	Indication for cholecystectomy
60/M	83	17	Hypersplenism	Symptomatic
20/M	84.8	13	Recurrent SSC	Incidental
36/M	76.8	20.9	Hypersplenism	Symptomatic
19/F	69.2	29.3	Recurrent SSC	Symptomatic
22/F	75.5	24.5	Hypersplenism	Incidental
22/F	86	14	Hypersplenism	Symptomatic
17/M	71.8	27.4	Splenic abscess	Symptomatic
45/F	75.8	22.9	Recurrent SSC	Symptomatic
15/F	77.6	21.2	Recurrent SSC	Symptomatic
14/M	75.4	23.8	Hypersplenism	Incidental
16/M	86.5	12.3	Recurrent SSC	Incidental
28/M	78.7	21.3	Hypersplenism	Symptomatic
20/M	75.4	23.2	Recurrent SSC	Incidental
14/F	76.5	20	Splenic abscess	Incidental
12/F	92.3	6.2	Recurrent SSC	Symptomatic
25/F	68.7	30.2	Recurrent SSC	Symptomatic
30/F	67	33	Hypersplenism	Symptomatic

SSC=splenic sequestration crisis.

as with improved care there is increased life expectancy and so a likely chance of developing gallstone-related complications.¹⁰ To avoid these complications and operating on these patients on an emergency basis, we advocate elective cholecystectomy even for asymptomatic gallstones.¹⁰

Because of the increased incidence of cholelithiasis in patients with SCD, it is not uncommon that patients undergoing splenectomy are found to have gallstones which can be either symptomatic or asymptomatic. The question is, should cholecystectomy be performed at the same time as splenectomy in these patients? During their study on the risks of synchronous gastrointestinal or biliary surgery with splenectomy for hematological diseases, McAneny et al.²¹ found that synchronous gastrointestinal or biliary surgery with splenectomy increases the risk of intra-abdominal abscess and, therefore, should be avoided. Pappis et al.,²² on the other hand, advocate simultaneous splenectomy and cholecystectomy in patients with chronic hemolytic anemia and cholelithiasis.

In our study of a high-risk group of patients with SCD, we found that concomitant splenectomy and cholecystectomy did not significantly increase morbidity. In six of our patients (35.5%), concomitant cholecystectomy was done for asymptomatic gallstones discovered incidentally during abdominal ultrasound evaluation.

Based on our results, and because of a high incidence of cholelithiasis in patients with SCD, screening for gallstones via abdominal ultrasound should be done in

those undergoing splenectomy. If gallstones are discovered, even if asymptomatic, simultaneous cholecystectomy and splenectomy should be done. With good perioperative care, including close liaison between the surgeon, anesthesiologist and hematologist, concomitant cholecystectomy and splenectomy in patients with SCD can be performed safely.

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