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Original Research Article

Gall stones in children with sickle cell disease, management and outcomes

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ABSTRACT

Background: It is rare to encounter a gall bladder stones in the paediatric age group. In Saudi Arabia, sickle cell disease (SCD) is common with incidence reach to 19.7%in some provinces. Cholecystitis is responsible for significant morbidity in SCD patients. Laparoscopic cholecystectomy (LC) is now considered as the procedure of choice for treating such cases. Over a period of 3 years, a total of 21 children with sickle cell anemia (SCA) had cholecystectomy. We exclude 3 patients who had splenectomy and cholecystectomy in the same time, the remaining 18 patients (6 males and 12 females) had LC only. Their age ranged from 4 years to 13 years (mean of 7.45±1.94 years).
Methods: LC was the standard procedure performed in all the children in our study group. The mean duration of operation was 92.17±27.31 minutes (range from 62-167 minutes).
Results: The mean post-operative stay after operation was 2.167±0.5 days (range from 1-3 days). One patient required conversion to open cholecystectomy (OC) because of severe adhesions, there were no immediate postoperative complications in the other 17 patients of our study group.
Conclusions: In conclusion, elective LC in well prepared patients, is feasible and safe in children with SCA and should be the treatment of choice for both symptomatic and asymptomatic cholelithiasis. ERCP is a valuable diagnostic and therapeutic investigation both preoperatively and postoperatively.

Keywords: Management, Gallstones, Sickle cell

INTRODUCTION

Sickle cell anemia is an inherited disease characterized by the presence of an abnormal hemoglobin in red blood corpuscles (RBCs). The frequent sickling leads to hemolysis with subsequent anemia.[1] The onset of the disease starts as early as at around 2 years old, but its prevalence increases progressively with age reaching 50% around 22 years old.[2] The diagnosis of SCA is made by a positive sickling test and Hemoglobin electrophoresis.[3] The chronic hyperbilirubinemia, due to hemolysis in sickle cell disease (SCD), frequently leads to the formation of pigment gallstones (cholelithiasis).[4] Generally, cholelithiasis is rare in the pediatric age. But, however, it is incidence is increasing in areas such as the eastern province of Saudi Arabia where SCA is diagnosed in up to 19.7% of children.[5] This frequency increased from 8.7% in those less than 10 years of age to 36% in those 15 to 18 years of age.[6] Cholecystitis is responsible for high levels of morbidity in SCD patients, and elective cholecystectomy is, therefore, the treatment

approach recommended to prevent acute complications as biliary tract obstruction, infection and pancreatitis.[7-10]

Cholecystectomy can be done by laparotomy via right subcostal incision. However, in the era of minimal invasive surgery, laparoscopic cholecystectomy (LC) is now considered as the procedure of choice.[11] This study is an analysis of our experience with LC in children with SCA, the management protocol and follow up.

METHODS

This is a retrospective study, all children with SCA who underwent laparoscopic cholecystectomy were reviewed as regarded: age, sex, total hemoglobin (Hb) level, Hb S level, clinical symptoms and signs, detailed operative notes, duration of surgery, post-operative complications and total duration of hospital stay, sickling test and Hb electrophoresis are the standard laboratory investigations to diagnose SCD. Preoperatively, all patients with Hb<10 g/dl have to receive simple blood transfusion to increase the Hb level to above level of 10 g/dl and hemoglobin S level should be below 45%. They were transfused with packed red blood cells using this formula: 3 ml/kg (Hb desired minus Hb on admission). Intravenous fluid hydration will start the night before operation day, 1½ times their maintenance requirement. Prophylactic antibiotics for 3 doses were given to all the patients.

Our technique is that the patient is positioned with a 30-degree tilt of the right side with a small roll placed under the right loin, a nasogastric tube inserted by the anesthesiologist for deflation of the stomach and kept in place along the whole procedure. We use initially 3-ports technique first at the upper edge of the umbilicus the second in the left midclavicular line above the level of umbilicus, the third in the right midclavicular line at or below the level of umbilicus all ports are of 5mm size, a fourth port of 3 mm may be added for traction in some cases in which a large pendulous fundus of the gall bladder that falls and mask the area of Callot's triangle, abdominal insufflation is done with low flow rate and up to pressure 12 mmHg. First step is to explore the abdomen and the liver and gall bladder then holding the fundus and retract it superiorly to expose the area under the liver to access the cystic duct and artery then start to create a window behind the neck of the gall bladder and skeletonize the cystic duct and artery using the hook electrocautery or the harmonic dissector. The cystic duct is clipped 2 proximal and one distal then the gall bladder is detached from the liver and its bed is inspected for any bleeding which will be secured by electrocoagulation, the gall bladder then removed either through the epigastric port if small and can be contained by 5 mm port or using end-bag 10 mm fig, the wounds are closed in layers. In the surgery ward Postoperatively, patients were given of pethidine (1 mg/kg) as analgesia every 8 hours for one day and continue simple non-steroidal analgesic as PRN. Feeding started when the bowel regains its function until reach full feeding This study was conducted for a period of (3) years (January (2018) and December (2021)), there were (21) children with SCA had laparoscopic cholecystectomy.

Table 1: Patient descriptive data.

	Mean	Range	Male	female
Age (years)	7.45±1.94	4-13		
Sex			6	12
Total Haemoglobin (gm/dl)	10.18±0.766	9.2-12.1		
Duration of Surgery (minutes)	92.17±27.31	62-167		

Figure 1: Preop pt. with previous laparotomy for intussusception at infancy.

Figure 2: Immediate post op three port site technique.

Figure 3: Gallbladder after excision with small stones.

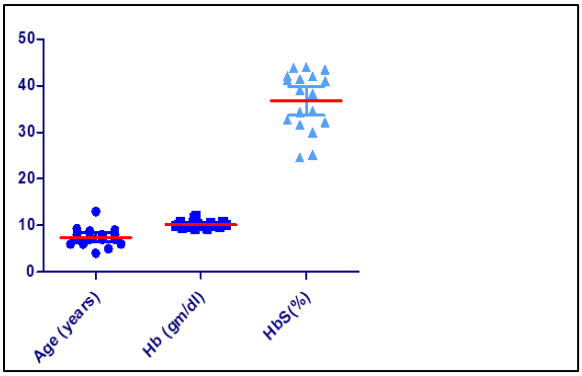

Figure 4: Mean age, hemoglobin (Hb gm /dl) and mean of hemoglobin S (Hb S %) after transfusion/ exchange transfusion preoperatively.

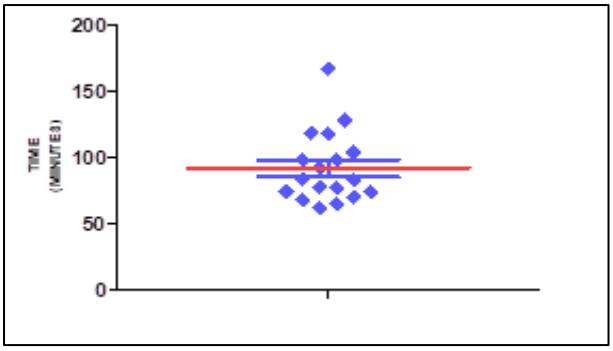

Figure 5: Depicting mean duration of operative time (minutes).

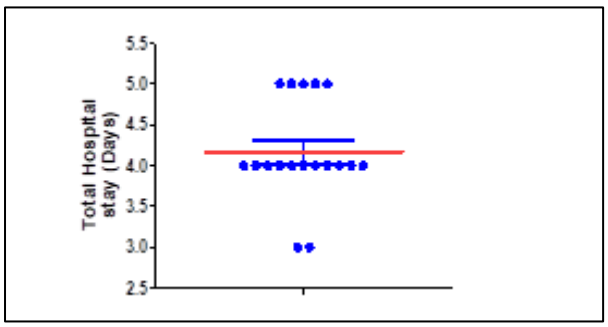

Figure 6: Depicting mean hospital stay (days) in our patients.

All these patients were investigated and prepared for surgery with target of Hbs less than 45 by simple blood transfusion or exchange transfusion. The most common indications for surgery were recurrent colicky abdominal pain with or without dyspepsia which was recorded in 11 (61.1%) patients, on and off nausea and vomiting in 6 (33.33%) patients, obstructive jaundice in 2 (11.11%) patients. 2 (11.11%) patients developed an episode of acute cholecystitis which was managed conservatively. 1(5.55%) patient developed acute pancreatitis, underwent ERCP and subsequent cholecystectomy in the same admission. 3(16.66%) patients were asymptomatic and had multiple gallstones discovered accidentally on ultrasound. Laparoscopic cholecystectomy was the standard procedure performed in all the children in our study group. The mean duration of operation was 92.17±27.31 minutes (range from 62-167 minutes). The mean hospital stay was 4.167±0.61 days (range from 3 to 5 days). One patient required conversion to OC because of severe adhesions, there were no immediate postoperative complications in the other 17 patients of our study group.

DISCUSSION

Since 1985 when Prof Dr Med Erich Muhe of Böblingen performed first LC. Then it has become popular as the procedure of choice to treat cholelithiasis both in children and adults.[12–15] In children with SCA, LC was shown to be a better option than OC.[16-18] It has advantages of better cosmetic, early recovery, shorter hospital stays, less post-operative pain and complications.[18] There is still controversy in the treatment of a symptomatic gall stones. Formerly, it was advocated to operate only if the patients with SCD have symptoms as pain, dyspepsia, or cholecystitis, to avoid postoperative complications which may reach up to 50% and mortality that may reach up to 10%.[19] Nowadays with improvement of perioperative care of those patients, surgery became safer. We consider operating on a symptomatic SCD patients to avoid the possible complications of gall stones including acute cholecystitis, pancreatitis, ascending cholangitis, and obstructive jaundice.[20,21] In vaso-occlusive crisis the abdominal pain may confuse with other emergency condition such as cholecystitis and appendicitis which will not be when patient already has cholecystectomy. [23]

In our patients, the mean hospital stay for LC was (42) days. This is relatively long, but the reason for this is that our patients are admitted 2 days before surgery for preoperative evaluation and some of them require a longer postoperative stay, but this is still shorter than the OC group.[23] In the general population with cholelithiasis, the incidence of CBD stones has been reported as 10% to 15%.10 In patients with SCA, the frequency of CBD stones is 18% to 30%.[7–9] The prevalence of choledocholithiasis in patients with SCA undergoing cholecystectomy as up to 30% was reported. Considering this high incidence, routine intraoperative cholangiogram was recommended in open cholecystectomy (OC). Laparoscopic CBD exploration is technically not easy although feasible the pediatric age group. It requires surgeon who are very experienced in laparoscopic surgery and it increases the time of surgery. Also, the hospital stay will be significantly increased with.[22–26]

In the era of laparoscopy and endoscopic retrograde cholangiopancreatography (ERCP), the question is whether laparoscopic cholangiography with CBD exploration is necessary in patients with SCA? ERCP has advantages of exploring the CBD and intrahepatic biliary radicles and treat the obstructive lesion either stones or strictures with minimal invasive approach.[23] The

indications for ERCP are those who have CBD dilatation or bile duct stones on ultrasound, a history of pancreatitis or ascending cholangitis. In these patients with normal ERCP, the magnetic resonance cholangio-pancreatography (MRCP) is now superior modality for detection of CBD stone in cases of obstructive jaundice with normal CBD on ultrasound in pediatric age group.[24–30] No patient requires CBD exploration when we start using ERCP pre-operatively. In conclusion, elective LC in well prepared patients, is feasible and safe in children with SCA and can be the best option for both symptomatic and asymptomatic gall bladder stones. Using ERCP and MRCP as a diagnostic and therapeutic investigatory tool in both preoperatively and postoperatively reduce the routine operative CBD exploration.

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