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Cholelithiasis in Infancy and Childhood: Spectrum and Current Management

MOSAD M. EL-BEHERY, M.D.; SAMI EL-SHIMI, M.D.
and AHMED EL-DORRY, M.D.

The Departments of Surgery, Pediatrics and Radiology, Faculty of Medicine, Ain Shams University.

Abstract

The advances of imaging studies have led to the recognition of cholelithiasis in infancy and childhood with increasing frequency. This study includes twenty eight children with cholelithiasis managed at the Surgical Departments of Ain Shams University Hospital, Cairo and Arabian Oil Company Hospital Saudi Arabia over the past 4 years. Their ages ranged from 1 to 12 years. Twelve children suffered from predisposing disorders: eight had gall stones in association with hereditary spherocytosis and underwent cholecystectomy during the course of splenectomy while four premature babies were treated with total parenteral nutrition (TPN) prior to the diagnosis of cholelithiasis and showed spontaneous stone resolution within 1 year. Sixteen children had idiopathic gall stones: twelve of them underwent open cholecystectomy (in addition to cyctogastrostomy for pancreatic pseudocyst in one of them), while four had laparoscopic cholecystectomy (LC). Follow up period ranged from 3 months to 4 years. We highlight the importance of nonoperative treatment of childhood cholelithiasis in selected patients aiming at spontaneous stone resolution and the advantages of applying laparoscopic cholecystectomy (LC) in children.

Introduction

CHOLELITHIASIS is relatively infrequent in infancy and childhood with an average prevalence between 0.13% [1] and 0.22% [2]. However, this disorder is being more recognized with improvement of imaging studies. Over the last two decades, the proportion of idiopathic gall

stones (i. e. without recognized aetiology) has increased up to 80% of childhood cholelithiasis while the incidence of stones resulting from hemolytic diseases has diminished to 20% or less [3&4]. Other risk factors include positive family history, total parenteral nutrition (TPN), extensive ileal resection, prolonged fasting and chronic illness such as cystic fibrosis [1]. This study reports the management and follow up of twenty eight children with cholelithiasis treated at Ain Shams University Hospital, Cairo and Arabian Oil Company Hospital, Saudi Arabia over the last 4 years.

Patients and Methods

Over the last 4 years, twenty eight children with cholelithiasis were managed at Ain Shams University Hospital, Cairo and Arabian Oil Company Hospital, Saudi Arabia. There were 18 males and 10 females. Their ages at the time of diagnosis ranged from 1 year to 12 years.

The children were divided into three groups according to the presence of potential risks of gall stone formation. The first group included 8 children with gall stones in association with hereditary spherocytosis, and second group (4 patients) were treated with TPN prior to diagnosis of cholelithiasis while the third group (16 patients) showed no specific risk factors (idiopathic gall stones). Each group was reviewed as regards age, sex, diagnostic modalities and lines of treatment. The follow up period ranged from 3 months to 4 years.

The data of the first group are presented in table (1). Patients showed no symptoms related to cholelithiasis, but the diagnosis was established by routine abdominal ultrasound before splenectomy. All patients showed radioopaque stones on plain x-ray. The 8 patients underwent cholecystectomy during the course of splenectomy. Chemical analysis showed that all stones are pigment ones.

The second group of patients included 4 children who were born prematures, and received TPN in the course of treatment of necrotizing enterocolitis at an age ranging from 2 to 4 months. Their data are presented in table (2). The children were asymptomatic with respect to gall bladder disease. The diagnosis of gall stones was initially made at an age between 1 & 2 years by ultrasound examination done for other pathology. The gall stones were radiolucent on plain x-ray. The 4 children were treated nonoperatively.

The data of third group are presented in table (3). All patients (except no. 4 & 12) presented with intermittent abdominal pain and tenderness, maximally felt in the right upper abdominal quadrant. Gall stones were diagnosed by ultrasound examination (Fig. 1 a,b) however, they were radiopaque in 3 patients only (no. 2,3&4). Patient no. 4 presented with acute cholecystitis and a gall bladder mass which proved to

Table (1): Data of Patients With Cholelithiasis and Hereditary Spherocytosis

Pt. No.	Sex	Age	Presentation	Diagnosis	Operation	Follow-up &Outcome
1	M	07 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	3.5 yrs Asymptomatic
2	M	5.5 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	2.5 yrs Asymptomatic
3	F	63/12 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	1.5 yrs Asymptomatic
4	M	4 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	1.5 yrs Asymptomatic
5	F	5 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	1.5 yrs. Asymptomatic
6	M	4 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	1 yr. Asymptomatic
7	M	5.7/12 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	1 yr. Asymptomatic
8	M	6 yrs.	Sibling of a father with hereditary spherocytosis Asymptomatic	US plain X-Ray	Simultaneous Splenectomy & Cholecystectomy	6 mon. Asymptomatic

be hydrops of the gall bladder (Fig. 2 a, b, c). Patient no. 12 presented with recurrent upper abdominal pain and epigastric mass, which proved to be a pancreatic pseudocyst by ultrasound and CT examination (Fig. 3 a,b). The first 11 patients of this group were treated with open cholecystectomy. In patient no. 12, a cystogastrostomy was added for the pancreatic pseudocyst, and laparoscopic cholecystectomy (LC) was performed for patients no. [13, 14, 15 & 15] with several technical modification [5 & 6].

Technique:

Under general anaesthesia, the child was placed in a supine Trendelenburg position. A nasogastric tube and a urinary catheter were placed. Pneumoperitoneum was obtained via a Veress needle introduced through a small incision just inferior to the umbilicus and in the 2 children it was sufficient to insufflate only 1.5-2 L. to keep the intraabdominal pressure less than 15 mm Hg. A 10-mm trocar was introduced followed by placement of the laparoscope with an attached video camera. Two lateral

Table (2): Data of the Patients with Cholelithiasis Following TPN Administration.

Pt. No.	Sex	Age	Presentation	Diagnosis	Treatment	Fate	Follow-up &Outcome
1	F	1 yr.	U.T.I.	US	Non-operative with U.S. follow-up	Spontaneous resolution after 1yr.	3 yrs. Asymptomatic
2	M	1.5 yrs.	Blunt abdominal trauma	US	Non-operative with U.S. follow-up	Spontaneous resolution after 9 months	2 yrs. Asymptomatic
3	M	2 yrs.	Blunt abdominal pain	US	Non-operative with U.S. follow-up	Spontaneous resolution after 6 months	1 yr. Asymptomatic
4	F	1.8/12 yrs.	U.T.I.	US	Non-operative with U.S. follow-up	Spontaneous resolution after 10 months	1 yr. Asymptomatic

Table (3): Data of the Patients with Idiopathic Call Stones.

Pt. No.	Sex	Age	Presentation	Diagnosis	Operation	Follow-up &Outcome
1	M	07 yrs.	Recurrent right upper abdominal pain	US	O/C	4 yrs. Asymptomatic
2	F	05 yrs.	Recurrent right upper abdominal pain	plain X-Ray US	O/C	3.5 yrs. Asymptomatic
3	M	03 yrs.	Recurrent right upper abdominal pain	plain X-Ray US	O/C	2.5 yrs. Asymptomatic
4	M	60 yrs.	Recurrent right upper &gall bdominal mass	Plain X-Ray U.S.& choles-cintigraphy	O/C	2 yrs. Asymptomatic
5	F	05 yrs.	Recurrent right upper abdominal pain	US	O/C	1.5 yrs. Asymptomatic
6	M	08 yrs.	Recurrent right upper abdominal pain	US	O/C	1.5 yrs. Asymptomatic
7	M	06 yrs.	Recurrent right upper abdominal pain	US	O/C	1.4/12 yrs. Asymptomatic
8	M	04 yrs.	Recurrent right upper abdominal pain	US	O/C	1.3/12 yrs. Asymptomatic
09	F	07 yrs.	Recurrent right upper abdominal pain	US	O/C	1 yr. Asymptomatic
10	F	06 yrs.	Recurrent right upper abdominal pain	US	O/C	1 yr. Asymptomatic
11	M	04 yrs.	Recurrent right upper abdominal pain	US	O/C	11months Asymptomatic
12	M	09 yrs.	Recurrent right upper & epigastric mass	US CT	Cystogastrostomy & Cholecystectomy	10months Asymptomatic
13	F	11 yrs.	Recurrent right upper abdominal pain	US	L/C	8 months Asymptomatic
14	M	10 yrs.	Recurrent right upper abdominal pain	US	L/C	6 months Asymptomatic
15	M	12 yrs.	Recurrent right upper abdominal pain	US	L/C	5 months Asymptomatic
16	F	10 yrs.	Recurrent right upper abdominal pain	US	L/C	3 months Asymptomatic

O/C : Open cholecystectomy L/C : Laparoscopic cholecystectomy

Fig. (1a & b): Ultrasound of the gall bladder showing gall stones (with acoustic shadow)

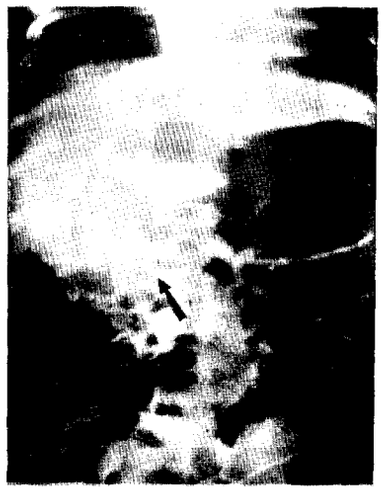
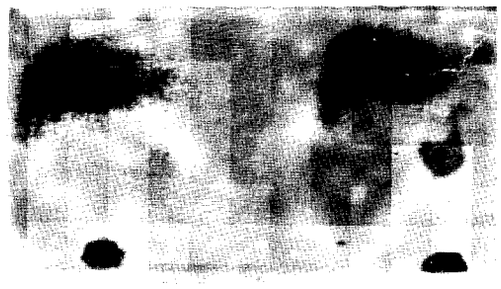
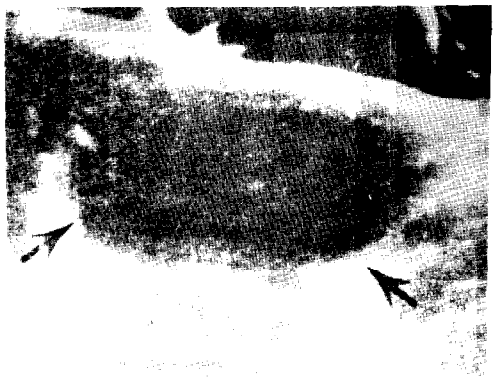


Fig. 2a: Plain abdominal X-Ray showing a gall bladder mass

2b: Ultrasound of the same patient demonstrating an evidence of cystic structure (gall bladder)



2c: Cholescintigraphy of the same patient showing absent accumulation of isotope in the gall bladder.



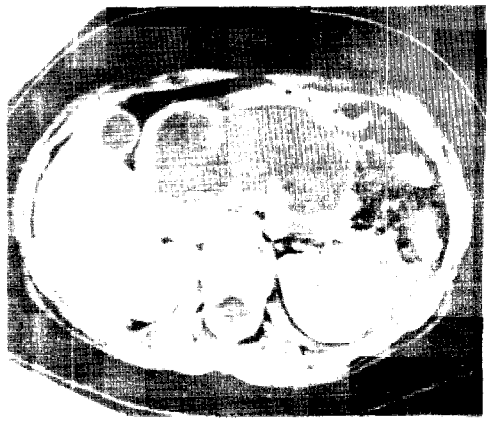


Fig. 3a: Ultrasound showing a pancreatic pseudocyst with some debris in the bottom. It is compressing the kidney below.

3b: CT of the same patient showing a pancreatic pseudocyst.

5-mm ports were put in the right midclavicular line & anterior axillary line 4-6 cm below the costal margin (ports 1& 2) and another 10-mm epigastric port (port 3) is placed 1 cm to the right of the midline half the distance from the xiphoid to the umbilicus (all these sites are more lateral and caudal than the usual adult sites to facilitate manipulation of gall bladder). Two grasping forceps were introduced through ports [1 & 2] to hold the fundus of the gall bladder and Hartmann's pouch in traction, while a dissecting forceps was introduced through the epigastric port. The cystic duct and artery were identified and skeletonised by removing the peritoneum overlying these structures. The cystic duct was isolated and transected between two proximal clips and distal one. The cystic artery was divided and secured with 2 clips proximally and a distant one. Intraoperative cholangiography was not performed 4 children as the cystic duct and the common bile duct were not dilated on US scan, the stones size was relatively big and there was no history of jaundice. The gall bladder was dissected from its bed using electrocautery. The laparoscope was then moved to port (3) and a grasping forceps was introduced through the umbilical port to extract the gall bladder. The gall bladder bed was irrigated with saline, the pneumoperitoneum was released and all the cannulae were removed. The umbilical fascia was closed with a vicryl suture and all skin incisions approx[illegible]ith subcuticular sutures. The nasogastric tube was

removed and the patient was transferred to the recovery room. The children were kept in hospital overnight, and a follow up outpatient examination was done 2 weeks after the procedure.

Results

The follow up period in this series ranged from 3 months to 4 years.

The 8 patients of the first group were followed up for 6 months to 3.5 years. They remain asymptomatic regarding the gall bladder disease (Table 1).

The 4 patients of the second group showed spontaneous stone resolution on US examination within 1 year after diagnosis. They are still asymptomatic after a follow up period of 1 to 3 years (Table 2).

On reviewing the 16 patients of the third group after follow up periods of 3 months to 4 years (Table 3). Fifteen had a smooth postoperative period and presently asymptomatic, while the patient who had cystogastrostomy suffered discomfort in the immediate postoperative period which settled on week after the surgery.

Discussion

Although once considered rare in infancy and childhood, cholelithiasis is currently being recognized with increasing frequency. This is due to the advent of ultrasound to detect asymptomatic gall stones which used to pass unrecognized. Also, the I. C. U. support to prematures and infants requiring TPN, added a new group of patients to the disease profile by improving their lift expectancy [7].

In our series, the idiopathic stones have been the majority (16 out of 28, i. e. 57.1%) compared to 80% in other series [3 & 4]. This reflects the current spectrum of the disease. Two risk factors for gall stone formation could be detected in our patients, viz., hereditary spherocytosis and TPN. Hereditary spherocytosis was found in 8 out of 28 patients (28.6%) compared to 20% in other series [3 & 4]. Other authors support our data of the predominance of hereditary spherocytosis over other blood diseases as a risk factor for cholelithiasis formation. Iwai et al. [8] reported associated gall stones in 43 - 63% of children with hereditary spherocytosis and 10-37% in sickle cell anemia, while Chittmittrapap et al [9] reported 2.3 - 23% of children with Thalassemia major.

Several reports have observed the association of TPN administration and gall stones with an incidence between 35 to 44% [10 & 11]. The mechanism is unclear, but may be related to alterations in bile from the amino acid infusions, lack of enteral feeding with reduction of enterohepatic circulation of bile and gastrointestinal dysfunction [12]. This association was found in 4 children in our series (14.3%) who had another contributing factor (prematurity) with decreased bile acid

output and shortened red blood cell life & increased bilirubin excretion [13].

In our series, available tests for diagnosis included plain abdominal radiography, ultrasonography, cholescintigraphy and CT scan. It was reported that 36-46% of gall stones in children were radioopaque [14], compared to 39.3% in our patients. However, we still recommend performing plain abdominal radiography to diagnose opaque stones which are less likely to undergo spontaneous resolution. In view of the relatively high frequency of asymptomatic gall stones in our series (12 out of 28, i. e. 42.9%), it is our recommendation to perform real time ultrasonography for children having risk factors of gall stone formation, e. g. hemolytic anemia and prolonged TPN administration. Also it should be a routine investigation for children with recurrent upper abdominal pain to detect possible radioopaque stones. The accuracy of ultrasound to detect gall bladder calculi is reported to be 96% [15], compared to 100% in our series. Although not commonly used to diagnose gall stones, CT scan was performed in one child to diagnose a pancreatic pseudocyst complicating gall stones. We also performed cholescintigraphy using 99m. technetium labelled iminodiacetic acid (IDA) in another child to confirm the presence of acute cholecystitis, being the most sensitive test for that purpose with a 100% accuracy rate [16].

There is still controversy regarding the best policy to treat cholelithiasis in children. Our policy is to follow a non operative treatment for a selected group of children with asymptomatic, non calcified gall stones with careful follow up by ultrasound to detect possible spontaneous resolution. This occurred in 4 children in our series. Other authors advised the same regime with a 12-month observation period [17 & 18]. On the other hand, we recommend cholecystectomy for asymptomatic calcified gall stones as no report documented resolution of that type of stones. We also recommend cholecystectomy for symptomatic gall stones or when complicated with acute inflammation or pancreatitis with pseudocyst formation or when gall stones are associating hemolytic anemia during the course of splenectomy. Some surgeons advocate cholecystostomy with simple removal of stones in the absence of inflammation of the gall bladder wall, and applied this technique in hemolytic diseases during the time of splenectomy [14]. In view of the reported recurrence of gall stones after cholecystostomy [3 , 19] we recommend performing elective cholecystectomy. This operation is tolerated well by most children and carries a very low risk. Other therapeutic options to treat gall stones in adults as oral chenodeoxycholic acid [20] and the use of concomitant extracorporeal shock wave lithotripsy [21,22] are not applicable in infants and children, due to the small percentage of patients qualified for this treatment, the long duration of therapy and the 50% risk of

recurrent stones in 5years [7]. Percutaneous puncture of the gall bladder or intrahepatic ducts with subsequent irrigation of the biliary sludge and stones was described in 10 infants and neonates by Parient et al. [23]. However, 4 patients of that series had cholangitis, comprising a high morbidity rate.

The standard method of gall bladder removal has been the open cholecystectomy through a right subcostal or a right upper paramedian incision. Perhaps, the most recent contribution to the management of gall bladder disease is laparoscopic cholecystectomy (LC). Several authors reported the use of LC in children weighing as little as 20 kg [5, 6, 24]. The four children who underwent this procedure in our series had the same advantages already described in adults, viz., shorter hospitalization, less atelectasis and ileus, reduced emotional stress and quicker return to normal activity. The significance of these advantages are even greater in children in that the quick return to home and school is psychologically beneficial for both the child and parents. LC is especially valuable to treat asymptomatic gall stones in sickle cell patients whose management is controversial [5]. Some authors advocate routine operative cholangiography during LC to prevent common bile duct injury and enhance identification of common duct stones [6, 25]. In our series, however, we did not perform intraoperative cholangiography for the patients who underwent LC. Our concept coincides with other authors' recommendation [24, 26] to perform a cholangiography when preoperative indicators suggest a higher risk of common bile duct stones, viz.: jaundice, pancreatitis or a dilated common duct.

We stress the point that if bleeding or concern about bile duct injury prevent safe LC procedure, the operation should be converted to open cholecystectomy.

It is our recommendation that LC should be within the grasp of pediatric surgeons as it is rapidly becoming the procedure of choice for treating cholelithiasis.

Conclusion

The possibility of gall stone disease should be considered in children with upper abdominal pain especially if there are risk factors e. g. hemolytic blood disease, prematurity or TPN administration. Infants with radiolucent stones following TPN administration may undergo spontaneous resolution and need close follow up. Laparoscopic cholecystectomy can be applied for children having cholelithiasis safely with less physical and psychological trauma.

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