

Changing trends in the management of choledochal cysts in children in an Egyptian institution

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Introduction/purpose Choledochal cyst excision and biliary reconstruction is the treatment of choice for choledochal cysts. Biliary reconstruction is carried out using several methods. The two most popular procedures are Roux-en-Y hepaticojejunostomy and hepaticoduodenostomy (HD). Both techniques have their merits and drawbacks. The aim of this study was to detect the change in trends in the management of cases of choledochal cysts during the period from 1993 to 2005 compared with the period between 2006 and 2015.

Patients and methods The files of patients admitted in our institute during the period from 2006 to 2015 for the treatment of choledochal cysts were reviewed. The data of patient' demographics, pathology, presentation, investigations, surgery, and outcome were statistically analyzed. Data were compared with a previous study conducted at this institute a decade earlier.

Results Files of 25 patients were retrieved. Patients' demographics, pathology, and presentation were not statistically different between the two decades. There was a recent change in trend in management; all cases were operated using HD, except for those cases that required

very high hepatointestinal anastomosis. HD had a significantly shorter operative time. Operation outcome and long-term follow-up were comparable between the two decades.

Conclusion HD is the preferred option for biliary reconstruction in our institute. This was based on the shorter operative time and avoidance of intestinal anastomosis, and an early and late complication rate compared with Roux-en-Y hepaticojejunostomy. *Ann Pediatr Surg* 12:94–99 © 2016 Annals of Pediatric Surgery.

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Introduction

Choledochal cysts were first described by Vater in 1723 [1]. They are classified into five subtypes under the Todani classification, based on the anatomical site of the cystic dilatation [2]. The estimated incidence of the disease ranges between one in 100 000 and one in 150 000 [3], and the male-to-female ratio ranges from 1:3 to 1:4 [4–6].

The classic clinical picture is a triad of abdominal pain, jaundice, and an abdominal mass [7]. Many authors reported that this triad is uncommon [8–10]. Clinical presentation varies with age, and 80% of the patients present before the age of 10 years.

The risk for developing biliary duct cancer in patients with choledochal cysts under the age of 10 years is 0.7%, which increases to 14.3% for patients over 20 years of age in western countries [11].

The treatment of choice is total cyst excision and biliary reconstruction. Cystoenterostomy without total excision is associated with an increased risk for cholangitis [12], pancreatitis [13], and biliary tract cancer [14,15].

The two main options for biliary reconstruction after cyst excision are Roux-en-Y hepaticojejunostomy (RYHJ) and hepaticoduodenostomy (HD), carried out either by open or laparoscopic approach [16,17]. Other surgical solutions are also available, such as jejunal interposition HD (the Chicago–Beijing procedure) and appendix interposition [18,19].

Postoperative complications, including postoperative cholangitis and intrahepatic stone formation, are due to anastomotic stricture and/or cholelithiasis [20]. The reported incidence rate of anastomotic stricture is 4.1% [21].

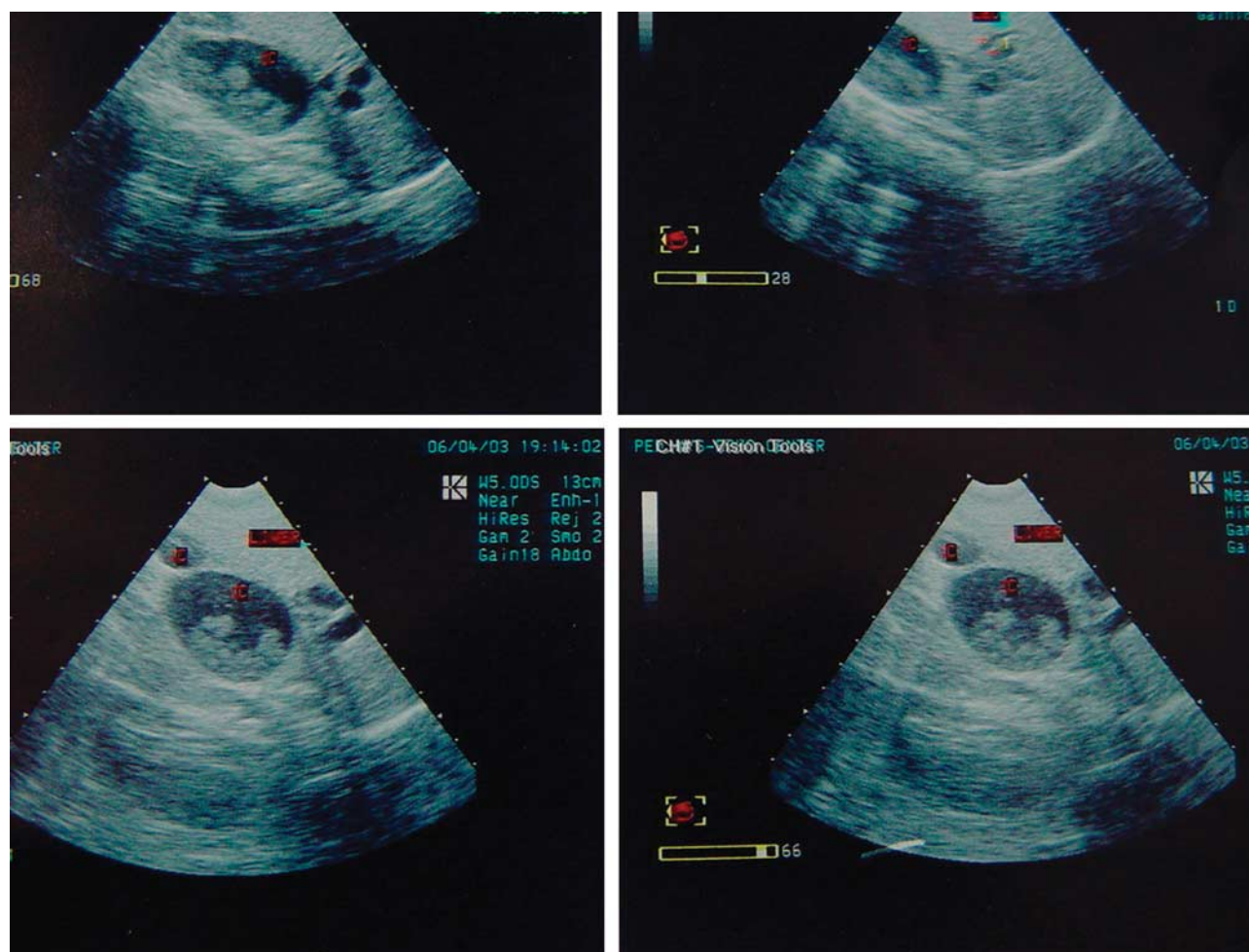
Total excision of cysts reduces the risk for cancer development. However, sporadic cancer has been reported a long time after the excision of cysts. Intrahepatic cholangiocarcinoma developing after total resection of choledochal cysts after 10–34 years has been previously reported [22–24].

In a previous study at this institution [25], the researchers reported their experience on this surgical condition, regarding the management and outcome. This study aimed at reporting the changes in the trends in managing these cases in our institute after that study.

Patients and methods

This retrospective study included patients with type I choledochal cysts who were admitted in the period from 2006 to 2015. Special charts were designed to retrieve the following data from the records: age at presentation, sex, type of the cyst, clinical presentation, investigations, surgical management modality, operative time, technical difficulties in reconstruction of the biliary tract, intraoperative and postoperative complications, and outcome of treatment. Follow-up data for patients were obtained by reviewing outpatient medical records, telephone interviews, and follow-up visits.

Fig. 1



Ultrasonography for a choledochal cyst.

Statistical analysis

Results were expressed as mean and SD. The comparison was made between the patients' demographics, investigations, operative decisions, and outcomes in our institute in the period 1993–2005 [25] and the period between 2006 and 2015 (this study). SPSS (version 16; SPSS Inc., Chicago, Illinois, USA) statistical package was used for data analysis. The Mann–Whitney *U*-test was used to compare quantitative data (a nonparametric test was used due to small size sample) and Fisher's exact test was used to compare qualitative data. A *P* value of less than 0.05 was considered significant.

This study was approved by our IRB. All patients parents were informed about the study, and an informed signed consent was obtained from all of them.

Results

Twenty-five cases were retrieved for the study. There were 17 (68%) girls and eight (32%) boys. The mean age at presentation was 14 ± 3.2 months.

The clinical presentation was jaundice (17 cases, 68%), abdominal mass with jaundice (six cases, 24%), and abdominal mass without jaundice (two cases, 8%).

Ultrasonography was diagnostic in 19 cases (Fig. 1). magnetic resonance cholangiopancreatography (MRCP) was carried out for six doubtful cases to ascertain the diagnosis (Fig. 2).

The cyst diameter ranged from 12 to 190 mm (Fig. 3), with a mean of 86.4 ± 9.68 mm. Total cyst excision was feasible in 24 cases, and intramural excision was done for one case (Fig. 4) in which marked adhesions due to inflammation made total excision impossible (Table 1).

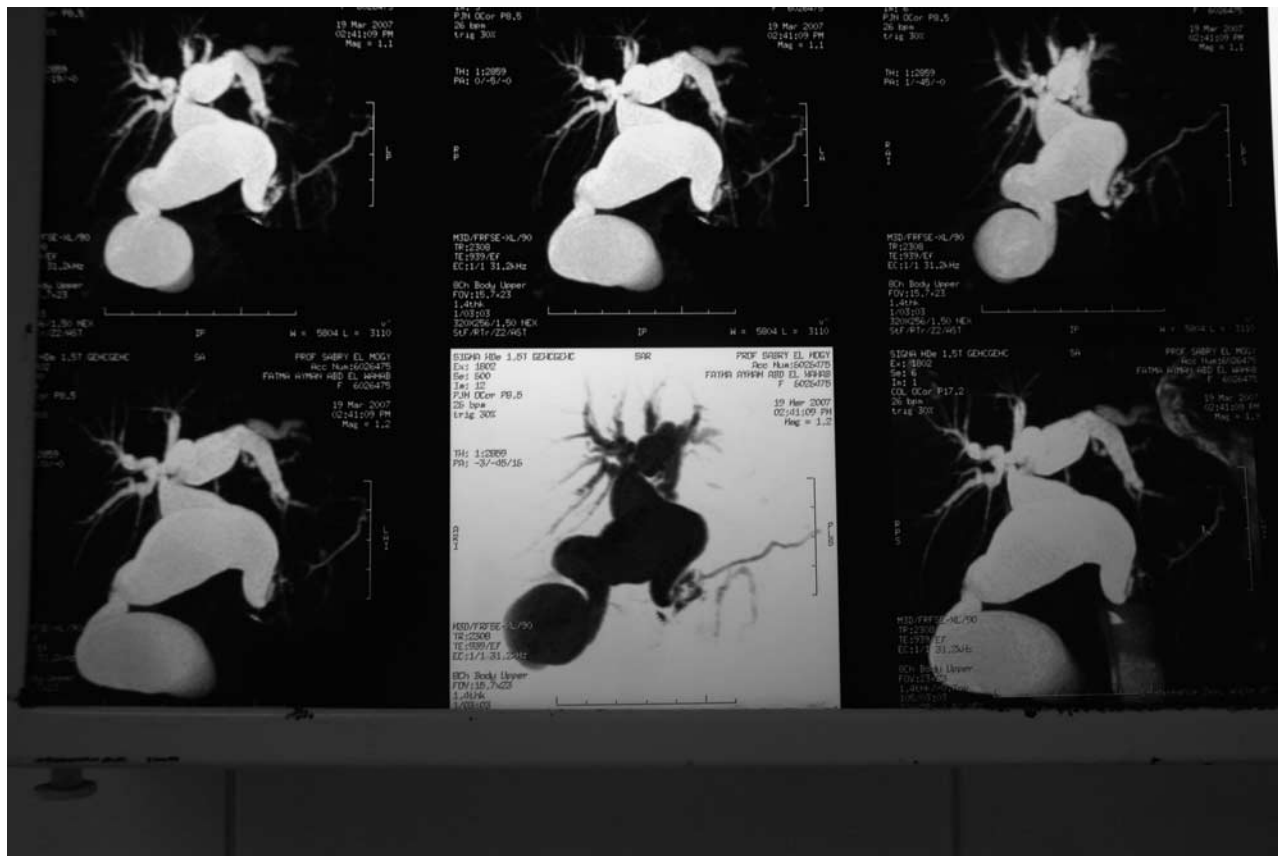
The standard approach in biliary tract reconstruction after cyst excision was HD (21 cases) (Fig. 5). RYHJ was reserved only for cases where anastomosis had to be done very high at the porta hepatis due to excision of the common bile duct and the common hepatic duct (four cases) (Fig. 6).

The operative time was 95 ± 5.1 min for HD and 160 ± 3.2 min for RYHJ. There were no recorded intraoperative complications.

Jaundice decreased gradually and clinically disappeared in all cases.

Prolonged ileus occurred in two case (had RYHJ), and responded well to conservative management. The two cases had wound infection, which was treated medically

Fig. 2



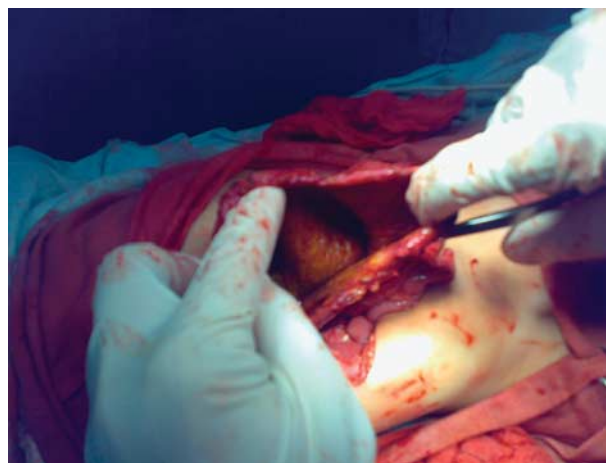
MRCP for a choledochal cyst.

Fig. 3



A very large choledochal cyst with a diameter almost 180 mm.

Fig. 4



Intramural dissection of a large cyst.

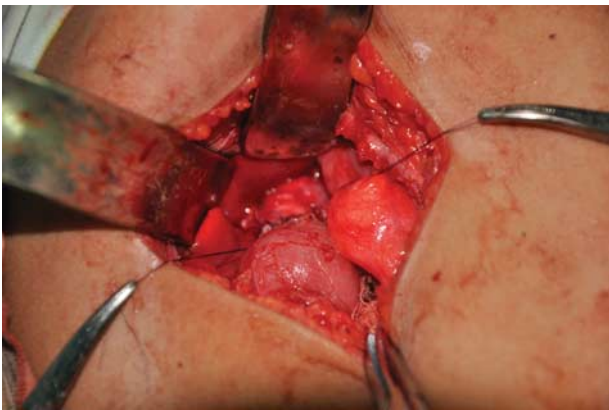
with success. Postoperative cholangitis occurred in one patient (HD reconstruction). Adhesive bowel obstruction occurred in two children (had RYHJ reconstruction), which was treated successfully by conservative management. All postoperative complications were not statistically different from those of the previous study.

The length of hospital stay was 5.34 ± 2.65 days, which was not statistically different from that of the previous study.

The length of follow-up was 54.6 ± 6.42 months. Long-term follow-up did not reveal any case of intrahepatic stone formation, or cases with biliary cancers (Table 2).

Table 1 Patients' demographics and clinical presentation

	Elhalaby <i>et al.</i> [25]	This study	<i>P</i> value
Age at admission (months)	12.35 ± 5.3	14 ± 3.2	0.427 (NS)
Sex			
Female	19	17	0.645 (NS)
Male	8	8	
Clinical presentation			
Jaundice	13	17	0.523 (NS)
Abdominal mass	3	2	
Mass + jaundice	11	6	
Cyst diameter (mm)	85.2 ± 12.68 (35–170)	86.4 ± 9.68 (12–190)	0.243 (NS)

Fig. 5

Hepaticoduodenostomy, showing the anastomosis at the porta hepatis.

Fig. 6

Roux-en-Y limb through a window in the transverse colon mesentery to the liver hilum.

Discussion

Surgical treatment of extrahepatic choledochal cysts has evolved over time. Before 1980, cystoduodenostomy and cystojejunostomy of Roux-Y were the popular procedures for choledochus cyst [18]. From 1981 through 1983, cyst excision plus hepaticojejunostomy of Roux-Y with a spur valve at the ascending limb was practiced [26]. In 1984,

the Chicago–Beijing procedure was introduced. It involves cyst excision plus a short jejunal interposition between the hepatic duct stump and the side wall of the descending duodenum with a spur valve at the duodenal anastomosis [27].

The most popular surgical approaches for biliary reconstruction after cyst excision are RYHJ and HD [28,29].

HD is preferred by some surgeons because it is simple to perform, has fewer complications such as anastomotic leakage, postoperative adhesive bowel obstruction, and peptic ulcer. Delivery of bile into the duodenum rather than into a Roux-en-Y (RY) limb of the jejunum is considered to be more physiological and is another advantage of using HD [20,30].

HD, however, is not preferred as a method for biliary reconstruction in cases of intrahepatic biliary duct dilatation or when the diameter of the common hepatic duct is more than 10 mm, due to reflux of duodenal contents into the intrahepatic ducts causing damage to its mucosa and cholangitis, anastomotic stricture formation, or even malignant transformation on the long run [31]. Another problem in HD reconstruction is duodenogastric bile reflux, associated with either gastritis or esophagitis [32].

RYHJ is a preferred approach by some surgeons [32]. Shimotakahara *et al.* [31] emphasized that the incidence of postoperative complications related to RYHJ, such as adhesive bowel obstruction and cholangitis (7.1%), is significantly lower than that of those related to HD, such as bilious gastritis (33.3%).

However, complications related to RYHJ are not uncommon [33]. They include stone formation in the elongated blind pouch (BP), intrahepatic bile duct stones caused by bile stasis in the elongated BP, bowel obstruction caused by adhesions between the elongated BP and small bowel, duodenal obstruction caused by compression by a tight RY limb vascular arch, and bile stasis caused by an abnormally elongated RY jejunal limb causing stone formation in the RY jejunal limb or intrahepatic ducts [33–35].

Stricture at the anastomosis is always the main factor of postoperative biliary infection. Stricture causes biliary stasis and stone formation. Reflux may induce food particles to be the core of a stone, and may also induce infectious organism. But if there is no stricture, the refluxed material may come out spontaneously [36]. To prevent the formation of anastomotic strictures, some

Table 2 Operative and outcome

	Ehalaby <i>et al.</i> [25]	This study	P value
Selection criteria for surgical approach	None	RYHJ reserved for near hilar anastomosis	
Reconstruction method			
HD	9	21	0.0001 (S)
RYHJ	18	3	
Operative time (HD and RYHJ) (min)	135 ± 3.2	98 ± 5.1	0.023 (S)
Cyst excision			
Total	24	24	0.611 (NS)
Intramural	3	1	
Prolonged ileus	4	2	0.669 (NS)
Postoperative cholangitis	4	1	0.352 (NS)
Adhesive bowel obstruction	1	2	0.603 (NS)
Length of hospital stay (days)	4.95 ± 1.76	5.34 ± 2.65	0.672 (NS)
Follow-up period	48.39 ± 4.7	54.6 ± 6.42	0.465 (NS)

HD, hepaticoduodenostomy; RYHJ, Roux-en-Y hepaticojejunostomy; S, significant.

surgeons recently have performed hepaticoenterostomy at the hepatic hilum, with a wide stoma [37].

In the previous study at this institute [25], the authors concluded that the outcomes of the two approaches are comparable, with significant short operative time in the HD group. This was reflected in our study. All cases after this study had HD reconstruction, reserving RYHJ to cases with involvement of the whole length of the common hepatic duct, making the anastomosis done nearly at the liver hilum; thus, HD becomes technically very difficult. Therefore, only three (12%) cases in this study underwent RYHJ reconstruction compared with 18 (66.67%) cases in the previous one.

The patients' demographics and clinical presentation were not statistically different between the two decades.

Diagnosis was confirmed in doubtful cases by using MRCP. No computed tomography was ordered for any of the cases in this series (compared with 29.6% in the previous study).

The operative time was significantly shorter in this study than the mean operative time in the previous one, because most cases underwent HD reconstruction, which takes lesser time to establish.

The difference in postoperative complications in both studies was statistically not significant. Likewise, the long-term complications, such as intrahepatic stones or carcinoma, were not encountered in both studies, perhaps due to the small number of cases in both.

Although choledochal cyst excision can be done laparoscopically [16,17], and this approach is practiced in some Egyptian institutes [38,39], it is still done exclusively via the open approach in our institute.

Conclusion

HD is the preferred option for biliary reconstruction in our institute. This was based on the shorter operative time and avoidance of intestinal anastomosis, and an early and late complication rate compared with RYHJ.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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