Surgical outcome of choledochal cysts in adults: a prospective cohort study

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Background
Choledochal cysts (CCs) are cystic dilatation of the biliary ductal system. Adult cases comprise around 20% of all cases.
Purpose
The aim of this study was to integrate all possible technical methods to prevent complications arising from residual choledochal tissue by presenting our experience in adult patients.
Patients and methods
A prospective cohort study of 24 adult patients, who underwent surgery for CC, over a 4-year period from March 2013 to February 2017 at the Gastroenterology Surgical Unit, Faculty of Medicine, Alexandria University, Egypt, was carried out. Cysts were classified according to the Todani classification. Biliary anatomy was defined by intraoperative cholangiography.
Results
The present study included six males and 18 females. Their ages ranged from 18 to 43 years (mean 26.4 years). Pain was the most common symptom at presentation (20 patients, 83.3%). Eighteen patients (75%) had type I cysts. All patients underwent excision of the extrahepatic bile duct cyst. The mean follow-up period was 34.6 months.
Conclusion
The present study showed satisfactory medium-term results following surgical resection of adult CC. Our approach was effective, to a great extent, in preventing complications of residual cysts. Excision of the extrahepatic bile duct should be guided by intraoperative cholangiography and distal clips to avoid pancreatic duct injury.

Keywords:
adult choledochal cyst, hepaticojejunostomy, intraoperative cholangiography, mucosectomy

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Introduction

Choledochal cysts (CCs) are cystic dilatation of the biliary ductal system, which can be extrahepatic, intrahepatic, or both. The disease is usually diagnosed in the first decade of life; however, adults comprise a substantial number of cases [1]. These patients may initially be asymptomatic, with subclinical bile duct inflammation, but later they are susceptible to recurrent abdominal pain associated with cholangitis, cholelithiasis, pancreatitis, or biliary obstruction [2]. The increased incidence of associated biliary tract stone disease, stricture formation, and superimposed malignancy lead to challenges in management of the disease [3].

There is a well-known, long-term association between CCs and cholangiocarcinoma. The risk increases with age. The risk in children younger than 10 years is less than 1%, rises to 18% in adults, and becomes greater than 50% in patients over 50 years. On the basis of Asian literature, malignancy occurs more often in types I and IV cysts and rarely in types II and III [4].

Complete cystic excision with cholecystectomy is the standard surgical approach as it lowers the risk of complications [5,6]. Types I, II, and IVb can usually be managed in this manner. Superimposed malignancy necessitates more extensive surgery, such as Whipple's operation for distal cholangiocarcinoma or hepatectomy for proximal cancers. Operations for adult choleduchal cyst are more difficult than their pediatrics counterparts because of the concomitant problems of inflammatory adhesions, infection, stone disease, malignancy, and previous surgery [3].

The diseased mucosa of the residual cyst may be the cause for postoperative complications. For that reason, mucosectomy of the cyst wall was advocated to prevent such complications in type IVa cysts [7–9].

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The literature is deficient in terms of studies on complete eradication of adult CCs and most series are retrospective in nature. Thus, we aimed to integrate all possible technical methods to prevent occurrence of long-term complications arising from residual choledochal tissue by presenting our experience in these patients.

Patients and methods

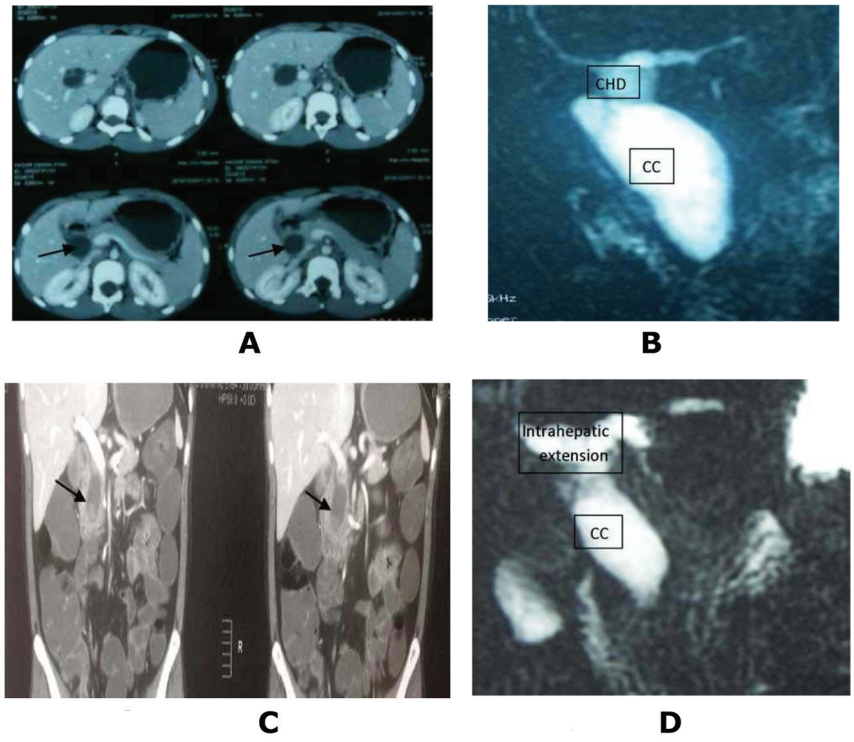
A prospective cohort study was designed including 24 adult patients who underwent surgery for CC during a 4-year period from March 2013 to February 2017 at the Gastroenterology Surgical Unit, Faculty of Medicine, Alexandria University, Egypt. Our center is the tertiary referral center of four major governorates (Alexandria, Beheira, Matrouh, and Kafr El-Sheikh). After obtaining approval of the local ethics committees, all patients were informed about the operative technique, and written consent was obtained. Patients were subjected to complete imaging starting with abdominal ultrasound; identified cysts were further delineated by both computed tomography scan and magnetic resonance cholangiopancreatography to estimate the size and extent of the disease and to assess for malignancy (Fig. 1). Endoscopic retrograde cholangiopancreatography was not included in the study protocol. Cysts were classified according to the Todani modification of the Alonso-Lej classification [10].

All patients received 1 g of third-generation cephalosporin and metronidazole at the time of induction of general anesthesia and then every 12 h for 5 days. A right subcostal incision was made. After dissecting the gallbladder, the hepatic flexure of the colon was mobilized, and the duodenum was kocherized. Biliary anatomy was defined by intraoperative cholangiography through a 8-Fr tube catheter introduced through the cystic duct. The length of the common channel between the distal end of the common bile duct (CBD) and the pancreatic duct, abnormal junction, or abrupt obstruction was recorded.

For types I and IVa, the choledochal dilatation is then dissected in an extramural plane between the peritoneum and the anterior wall. Dissection progressed across the lateral walls downwards, separating the cyst from the first part of the duodenum (Fig. 2).

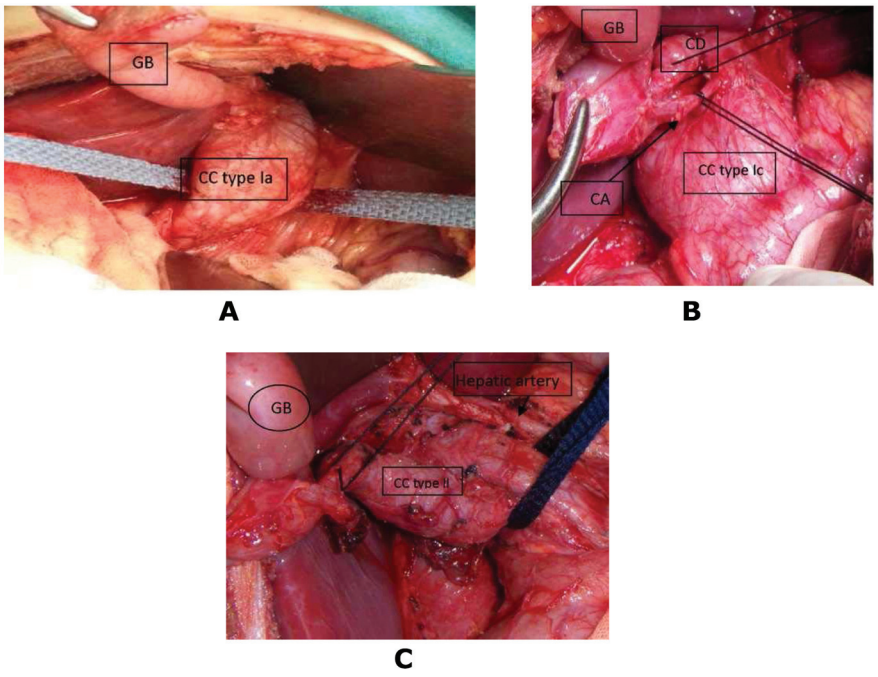
Next, a clip was tucked to the inspected distal limit of the CC, and a repeat cholangiography was carried out to check the accurate lower limit. If the cholangiogram showed distal extension of the cyst, the dissection continued further downwards (Fig. 3).

Figure 1



(a) Computed tomography scan of the abdomen showing type Ia choledochal cyst (black arrow). (b) Magnetic resonance cholangiopancreatography (MRCP) showing type Ia choledochal cyst (CC). (c) Coronal section showing type Ic choledochal cyst harbouring intracystic stone (black arrow), (d) MRCP showing type IVa choledochal cyst

Figure 2



(a) Type Ia CC involving extrahepatic bile duct. (b) Type Ic CC. (C) Type II CC. CA, cystic artery; CC, choledochal cyst; CD: cystic duct; GB, gallbladder

The distal bile duct was then over-sewed by propylene 3/0 sutures and transected just above the demarcated pancreatic duct within the head of the pancreas, to avoid leaving any intra-pancreatic part of the cyst (Fig. 4).

In case of difficulties caused by the close posterior proximity and adhesions of CC to adjacent structures in the hepatoduodenal ligament (mainly to the portal vein), the anterior cyst wall was incised allowing the technique of mucosectomy, formerly published by Lilly's, to be adopted (Fig. 5).

The proximal part was dissected until the level of the hilar bifurcation and then transected. Proximal intrahepatic ducts were cleared from debris by irrigation. If the mucosa of the residual intra-hepatic cyst wall was found to be inflamed and thickened, the mucosa was excised or stripped off like a sleeve. Bleeding from the exposed bile duct wall was controlled with bipolar diathermy or by compression.

A 40-cm retrocolic jejunal Roux loop was used to construct the end-to-side bilioenteric anastomosis using interrupted absorbable sutures (4/0 polyglactin) with accurate apposition. The anteroinferior wall of the extrahepatic segment of the left hepatic duct was incised for a spacious hepaticojejunal anastomosis. In cases where a narrow anastomosis was expected, a 10-Fr plastic biliary stent was inserted to prevent early stricture formation.

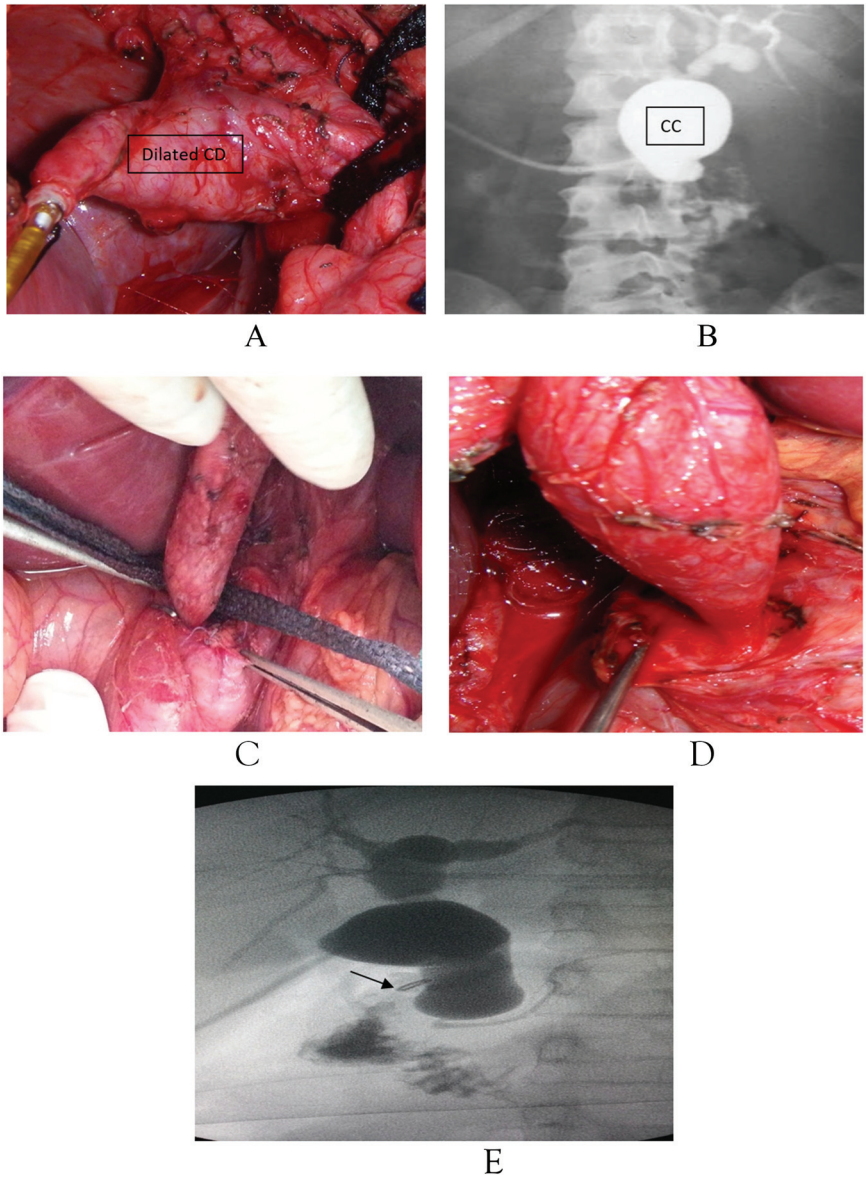
An abdominal drain was inserted in all cases. All resected specimens were sent for histopathological examination (Fig. 6). Patients resumed oral fluid intake after 3–5 days and then a soft diet when tolerated before discharge.

Operative/postoperative findings, pathology results, and follow-up outcomes were collected. Early and late postoperative complications were noted. All patients were seen in the outpatient clinic at 1 week, 1 month, and 3 months postoperatively, and at 6-month intervals thereafter. Data are presented using numbers, percentages, arithmetic means, and SD, and were analyzed using SPSS (Chicago, IL, USA) (version 15) statistical software.

Results

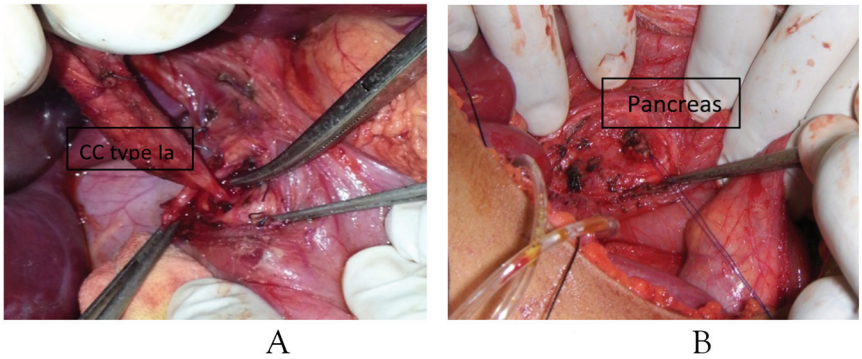
Twenty-nine adult patients with CC presented to us during the study period; three of them were excluded because of confirmed diagnosis of cholangiocarcinoma at the time of presentation. Two patients with Caroli disease were excluded as well because their treatment was medical and liver transplantation, which is still not feasible at our institution. Thus, 24 adult patients who underwent surgery for CC were included. There were six males (25%) and 18 females (75%) with a

Figure 3



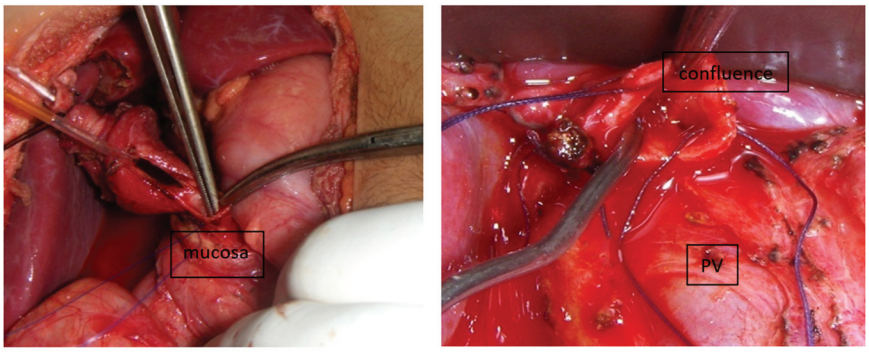
(a, b) Cystic duct catheterization and intraoperative cholangiography were performed, the cystic duct was dilated. (c, d) Operative image showing the distal clip tucked to the inspected lower level of the choledochal cyst (black arrow) and distal dissection was continued further (e) Operative cholangiogram of the same patient showing extension of type Ia choledochal cyst for 2 cm distal to clip tucked (black arrow) to the inspected lower level of the cyst

Figure 4



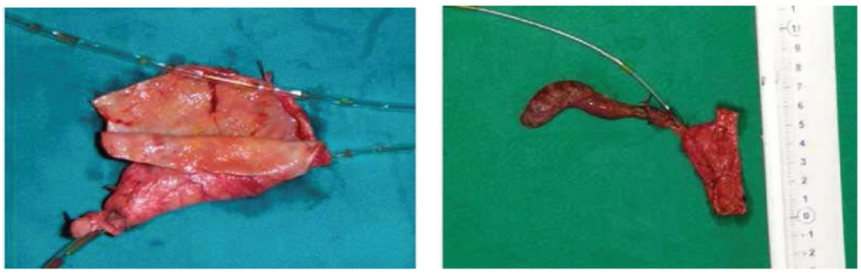
(a) Distal retroduodenal intrapancreatic dissection till normal diameter of common bile duct, checked by operative cholangiogram. (b) Suturing of lowermost level after transection of the choledochal cyst by prolene sutures away from pancreatic duct guided by intraoperative cholangiogram

Figure 5



(a) Distal mucosectomy of posterior wall of the cyst. (b) Opening of hepatic duct confluence after proximal transection of the cyst ready for hepaticojejunal anastomosis, the mucosa of the residual intrahepatic cyst wall was excised in piecemeal or stripped off (proximal mucosectomy)

Figure 6



(a, b) Resected specimens of type I choledochal cysts showing mucosal inflammation, thickening, and fibrosis

female-to-male ratio of 3 : 1. The age of patients at diagnosis ranged from 18 to 43 years (mean 26.4 years).

Epigastric dull aching pain was the most frequent symptom at presentation (20 patients, 83.3%) followed by nausea and vomiting (10 patients, 41.6%). One episode of cholangitis had been experienced by one-third of patients. Jaundice was found in seven patients (29%). A past history of pancreatitis was obtained from three patients (12.5%). The classic triad (abdominal pain, jaundice, and a palpable mass) was not seen in any patient. None of the patients had undergone previous surgery for CCs in their childhood.

On the basis of Todani's classification, 18 patients (75%) had type I CC (Figs 1 and 2), four cases (16.6%) had type IVa cysts (Fig. 1), and two cases (8.3%) had type II cysts (Fig. 2). In type IVa cysts (four cases), three cases were bilobar, and in one case it was confined to the left hepatic lobe. Regarding the presenting symptom compared with the type of cyst, abdominal pain was the main symptom in most types. Jaundice was seen in type I (four of 18 patients) and type IV (three of four patients) cysts.

Eight patients had biliary stones at presentation, including four cases with gallstones (16.6%), with three patients (12.5%) harboring stones in their extrahepatic CCs, and two of them (8.3%) having intrahepatic ductal stones (one patient had both intrahepatic and extrahepatic stones). In 17 patients (71%), abnormal pancreaticobiliary junction was demonstrated on intraoperative cholangiography. Sudden and severe narrowing denoting distal obstruction of terminal choledochus with normal pancreaticobiliary junction was seen in five (21%) patients. The clinical presentation, Todani classification, laboratory results, and morphological features are given in Table 1.

All patients underwent excision of the extrahepatic bile duct cyst (type I, II, and the extrahepatic part of type IVa) with reconstruction by a Roux en-Y hepaticojejunostomy in 22 patients. Three cases with type IVa cysts were not completely excised, although left hepatectomy was performed in one patient. There were two cases of type II cysts in our series – one of

Table 1 Clinical presentation, Todani's classification, laboratory results, and morphological features in the studied adults with choledochal cyst

Clinical presentation	*N* (%)/mean (range)
Abdominal pain	20 (83.3)
Nausea and vomiting	10 (41.6)
History of cholangitis	8 (34)
Jaundice	7 (29)
History of pancreatitis	3 (12.5)
Todani classification	
Type I	18 (75)
Ia	13 (72.2)
Ib	2 (11.1)
Ic	3 (16.6)
Type II	2 (8.3)
Type III	–
Type IVa	4 (16.6)
Type V	–
Laboratory results	
Serum albumin (g/dl)	3.48 (2.5–4.5)
Serum alkaline phosphatase (U/l)	304 (128–536)
Serum ALT (U/l)	63.58 (33–156)
Serum AST (U/l)	32.9 (13–58)
Serum bilirubin, total (mg/dl)	1.42 (0.3–5)
Morphological features	
APBJ	17 (71)
Distal obstruction	5 (21)
Bile sludge/stone formation	8 (33.3)
Accessory hepatic ducts	2

ABPJ, abnormal pancreaticobiliary junction; ALT, alanine aminotransferase; AST, aspartate aminotransferase.

them was managed by simple cyst excision and direct axial repair of the bile duct over a T-tube and the other by segmental resection of the CBD and choledochoduodenostomy.

Extensive adhesions with adjacent structures (mainly to portal vein) were noted in nine patients (37.5%), and in these cases a part of the cyst wall was left behind after excising the mucosa. The mean width of the hepaticojejunostomy was 40.5±6 mm (range 12–55 mm). The anstomosis was performed at the level of the carina or common hepatic duct (single duct anastomosis). The mean operating time was 185.3±46.9 min (range 120–240 min), and the mean operative blood loss was 175.3±50.7 ml (range 100–250 ml). The mean hospital stay was 11.3±4 days (range 5–21 days).

A subcutaneous hepaticojejunal access loop was made in one patient with type IVa having both intrahepatic and extrahepatic stones. The jejunal loop was marked by clipping the sutures holding the access loop in place.

As shown in Table 2, the total complication rate was 41.6% (10/24). Early complications were noted in six

Table 2 Distribution of patients according to early and late postoperative complications

Postoperative complication	*N* (%)
Early complications (*N*=6)	
Bile leak	1 (4.1)
Intra-abdominal collection	1 (4.1)
Wound infection	5 (20.8)
Wound hematoma	1 (4.1)
Total	8[a]
Late complications (*N*=7)	
Intrahepatic stones	1 (4.1)
Anastomotic stricture	3 (12.5)
Cholangitis	2 (8.3)
Pancreatitis	1 (4.1)
Incisional hernia	2 (8.3)
Total	9[a]

[a]Some patients had more than one complication.

patients (25%) including wound infection (five patients) and hematoma (one patient). A patient (4.1%) had biliary leak with subhepatic abscess, which resolved after 1 week of parenteral nutrition, intravenous antibiotics, and percutaneous drainage; enteral feeding was delayed for 10 days in this patient. One patient was found on postoperative ultrasound study to have minor fluid collection in the gallbladder fossa that resolved spontaneously on follow-up. No perioperative mortality occurred.

Seven patients (29.1%) had late postoperative complications: recurrent intrahepatic stones in one, anastomotic stricture in three, cholangitis in two, pancreatitis in one, and incisional hernia in two. Two patients developed cholangitis that resolved with medical treatment in one, and in the other patient percutaneous transhepatic drainage was performed. Three patients (two type I and one type IVa) developed anastomotic stricture. Two patients (type I) required percutaneous dilatation of the stricture and insertion of a biliary stent by the interventional radiologist. In the third patient, access to the biliary tree was achieved using the subcutaneous access limb. This patient also developed recurrent intrahepatic stones in the left lobe of the liver on top of the stricture. His condition was detected after 1.5 years. He had a type IVa cyst with intrahepatic and extrahepatic biliary stones. An end-viewing gastroscope was passed through the subcutaneous access limb (identified by clips on C-arm). After entering through the anastomosis, the hepatic ducts were reached, and a cholangiogram was performed. Dormia basket and extraction balloons were successfully used to extract calculi. A balloon catheter over a guide-wire was inserted through the working channel of the scope and then through the stricture

and inflated to 5 mm for 10 min to dilate the stricture. On completion, the ducts were flushed with saline, and the jejunal loop was closed.

Histopathological examination showed the presence of cyst wall inflammation in 22 (91.6%) and cholecystitis in 12 (50%) patients. Premalignant proliferative changes were detected in two cases.

On follow-up, patients were in an acceptable state of health. The mean follow-up duration was 34.6±13.2 months (median 1.5 years), ranging from 1 to 48 months. All patients were assessed by liver function tests and abdominal ultrasonography. No malignant disease was recorded.

Discussion

Bile duct cysts are one of the distinguished congenital disorders that cause dilatation mainly of the extrahepatic and sometimes the intrahepatic biliary tree. Despite being a rare pediatric disorder, we were able to find and manage 24 adults with CCs over a relatively short period. The number of cases of such a rare disease may be attributed to several factors: first, our center is a tertiary referral center serving four densely populated governorates. Second, the disease may be asymptomatic that adult presentation can exist in up to one-third of the cases of CCs [6–8]. Finally, symptoms are vague and nonspecific, and the low level of education and socioeconomic state of parents in some rural areas preclude early referral of their child for medical consultation till adulthood.

Abdominal pain was the most common complaint in this report (20 patients, 83.3%). It has been reported that the type of symptoms depends largely on the age at presentation – abdominal pain is the most frequent symptom in adults, whereas jaundice is the main presentation in children [8].

In this series, type I constituted the largest group, accounting for 75% of cases, followed by type IV (16.6%), similar to other studies [3,11–15]. In a North American series [16], type IVa was the predominant type, and the authors recommended routine intraoperative cholangiogram to reveal intrahepatic association in patients with type I.

Harsh difficulties were met during dissection in nine cases (37.5%). These surgical difficulties arose from severe adhesions to neighboring vital vessels, mainly the portal vein. Tsai *et al.* [15] reported that cyst excision was much more difficult in adults than their pediatric counterparts because of the presence of more intense pericystic inflammation with distortion of the anatomy and more bleeding during dissection. In practice, complete excision can be performed by careful dissection around the cyst towards the lowermost normal-diameter portion of the CBD with the help of traction, avoiding injury to the pancreatic duct. Incomplete excision results in retention of residual tissue that can give rise to complications such as stone formation, pancreatitis, and malignancy [3,6,7,11,13–19].

In the same way, we recommend proximal cyst resection till the ductal confluence, and reconstruction by an ample roux-en-Y wide hepaticojejunostomy. To prevent anastomotic strictures, many studies [6,16,20] recommended high, wide anastomosis. We recommend proximal anastomosis with a nonpathological part of common hepatic duct.

We encountered four patients (16.6%) with type IVa cysts. Type IVa cysts are recognized to be more common in adults with a reported prevalence ranging from 2 to 39% [21–23]. This wide range of type IVa cyst may be related to underestimation of the intrahepatic involvement. Therefore, accurate imaging of type I cysts in adults is of utmost importance. For these cases, the extrahepatic component was completely resected as for type I cyst. Unfortunately, three cases had diffuse cysts in both lobes of the liver that precluded liver resection, and therefore mucosectomy of intrahepatic ducts was performed. On follow-up magnetic resonance cholangiopancreatography, shrinkage of intrahepatic parts was identified in two cases. Huang *et al.* [14] and Dutta [7] stated that intrahepatic involvement in type IVa is confusing, and some of their patients showed regression of intrahepatic components after excision of extrahepatic CC and adequate drainage. In the same way, Koshinaga *et al.* [23] recognized that some barrel-shaped biliary dilatations vanished after total excision.

Excision of the inner layer of the cyst wall or mucosectomy provided a compromised solution for management of both hilar intrahepatic cysts and distal intrapancreatic extension. In this procedure, a plane was created between the pathological mucosa and the underlying wall of the cyst with difficulty. Curettage of the abnormal friable mucosa was also performed in some cases.

In the present study, anastomotic stricture developed in three patients after mucosectomy. We believe that ample-diameter hepaticojejunal anastomosis, with a mean of 40.5 mm, was crucial in lowering the risk of postoperative tight anastomosis. Cyst wall inflammation

in adults is intense, damaging the common hepatic duct that is used for hepaticojejunostomy [24]. Two patients with type I cyst underwent percutaneous balloon dilatation and insertion of a biliary stent. This concurs with other investigators who recommend this approach for anastomotic strictures [24,25].

Intraoperative cyst cholangiography allowed us to precisely excise the distal end of the cyst, without injuring the pancreatic duct as its opening can be identified, leaving only a minimal intrapancreatic terminal choledochus. During surgery, debris or protein plugs in the common channel were removed by irrigating normal saline. In agreement, Wiseman *et al.* [16] recommended the routine use of intraoperative cholangiogram to diagnose intrahepatic extension of the disease.

All possible surgical means were performed to avoid risks of remnant complications: distal clip-guided intraoperative cholangiography, mucosectomy at proximal, cystic, and distal levels, extensive intrapancreatic dissection, wide conduit, and the selective use of subcutaneous access limb in risky cases to aid further management.

Malignancy occurs in 2.5– 30% of patients with CC, and jaundice should raise suspicion of malignancy in adults [2,25–27]. Lee *et al.* [3] reported an incidence of malignancy of 20% in 25 adults. They suggested that a high degree of suspicion should be applied in older male patients and in the presence of impaired liver function. Pancreatic secretions regurgitating into the retained abnormal biliary tissue embedded in the pancreas may trigger malignant cells to proliferate [28].

In the present study, no malignancy has been detected in any of our cases until now. The mean age at presentation in this series was 26.4 years – an age with a relatively lower malignant risk than in those over 50 years. Delayed presentation without simultaneous cancer in an untreated adult warrants an eminent risk of future malignancy. Kobayashi *et al.* [29] reported that the epithelium of the retained bile duct may have already undergone genetic alterations to a premalignant stage at the time of surgery, and these genetic changes may continue during the postoperative period.

Two histopathological reports found premalignant changes in our series. Komi *et al.* [30] and Komuro *et al.* [31] highlighted increasing rates of premalignant changes in resected cysts with advancing age. Hence, long-term follow-up is considered. In this report, one patient had an episode of pancreatitis on follow-up. On the other hand, cholangitis was found in two cases; this may be explained by relative bile stasis and exposure to enteric milieu; 34% of adults in the retrospective series of Huang *et al.* [14] had severe late complications such as cholangitis, biliary cirrhosis, and strictures.

The small sample size from a single center, a medium-term follow-up period, and lack of feasibility to liver transplantation were noted limitations of this study.

Conclusion

The present study showed satisfactory medium-term results following surgical resection of adult CC. Our approach was effective, to a great extent, in preventing complications of residual cysts. Imaging is the cornerstone for the complete diagnosis and subsequent surgical management, notably for intrahepatic involvement. Excision of the mucosa of the intrahepatic cyst enables further resection of the affected mucosa with attendant risk of stricture. Excision of the extrahepatic bile duct should be guided by intraoperative cholangiography and distal ligaclips to avoid pancreatic duct injury. Long-term surveillance remains essential. Being an uncommon disease, larger-scale studies are required for more data and experience for optimum management.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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