Management of Choledochal Cysts In Gastroenterology and Hepatology Teaching Hospital

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ABSTRACT:

BACKGROUND:

A Choledochal cyst is a rare biliary disease mostly presenting during childhood. Adult presentation is rare and associated diseases and complications are common.

OBJECTIVE:

This study aims to review the management of patients who presented to our hospital with choledochal cysts, focusing on their presentation, preoperative investigations, treatment given and postoperative course.

METHODS:

A prospective and retrospective review of all our choledochal cysts patients from April 2000 to November 2010 was performed

RESULTS:

There were twenty three patients, sixteen females and seven males the average age was 25.82 (range 4-70) years. The commonest presenting complaints were abdominal pain or jaundice. There were eighteen Types I (78.2 %), two of the adult patients had concomitant cholangiocarcinoma (8.6 %), and four patients had cholangitis (two of them had associated cystolithiasis). Fourteen patients treated by total cyst excision with hepaticojejunostomy,

CONCLUSION:

Adult patients with choledochal cysts have associated biliary problems. In view of the high risk of cholangiocarcinoma, early resection and not internal drainage is the appropriate treatment.

KEYWORDS: choledochal cysts, cholangiocarcinoma, biliary diseases

INTRODUCTION:

Biliary cysts are cystic dilatations, which may occur singly or in multiples throughout the bile ducts. They were originally termed choledochal cysts (involving the extrahepatic bile duct) but the clinical classification was revised in 1977 to include intrahepatic cysts [1,2].The incidence has been estimated to be 1:100,000 to 150,000 In the past, the majority of cases were reported in children, although more recent series report equal numbers in adults and children [3].Pathologic features of choledochal cysts are variable, ranging from normal bile duct mucosa to carcinoma [4,5]. Evidence clearly points to a 20 to 30-fold increased risk of cholangiocarcinoma in biliary cysts compared to the general population [6].Several theories of biliary cyst formation have been proposed, and it seems likely that no one mechanism accounts for all biliary cysts. Cysts may be congenital [7]or acquired]. an abnormal pancreaticobiliary junction (APBJ), also called pancreaticobiliary maljunction (PBM), is a rare congenital anomaly, with prevalence of 0.03 percent

APBJ is characterized by junction of the bile duct and pancreatic duct outside the duodenal wall [8], is present in about 70 percent of patients with biliary cysts and may be a significant risk factor for the development of malignancy in the biliary cyst [9].

Infants with biliary cysts commonly present with conjugated hyperbilirubinemia (80 percent), failure to thrive, or an abdominal mass (30 to 60 percent). The triad of pain, jaundice, and abdominal mass is found in 11 to 63 percent [4]. In contrast, chronic and intermittent abdominal pain appears to be the most common presenting symptom (50 to 96 percent) in patients older than two. A diagnosis of biliary cyst should be considered in adults when a dilated portion of the bile ducts or ampulla is identified, especially in

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the absence of overt obstruction. Cross-sectional imaging with ultrasound or CT may suggest the presence of a biliary cyst. Direct cholangiography (whether intraoperative, percutaneous, or endoscopic) has long been considered the best test for diagnosis and evaluation.

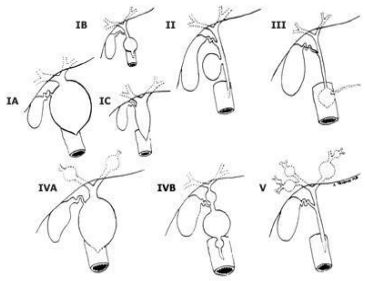
MRCP also appears to be useful for diagnosis. It accurately demonstrates cystically dilated segments of the biliary tree, and identifies APBJ in over 75 precent of cases [10].

However, MRI is less sensitive than direct cholangiography for excluding obstruction.

Endoscopic ultrasound (EUS) can also demonstrate extrahepatic biliary cysts and provide detailed images of the cyst and pancreaticobiliary junction.

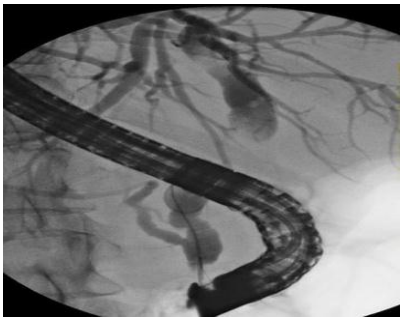
Because of the risk of malignant transformation of the cyst, the current standard of treatment for Type I, II, and IV biliary cysts is surgical excision, with the goal of removing all of the cyst tissue when possible.

Classification of choledochal cysts according to Todani and colleague



IA)common type; (IB)segmental dilatation; (IC)diffuse dilatation;(II) diverticulum; (III)choledochocele; (IVA)multiple cysts (intra- and extrahepatic);(IVB) multiple cysts (extrahepatic); (V) single or multiple dilatations of the intrahepatic ducts.

Anomalous pancreaticobiliary junction



Endoscopic retrograde cholangiopancreatography in an adult with obstructive jaundice demonstrates an anomalous pancreaticobiliary junction with a malignant biliary stricture replacing the cystic duct insertion. There is no evidence of a biliary cyst.

Type I biliary cysts

Image obtained after ERCP showing a type I biliary cysts associated with an anomalous pancreaticobiliary junction.

PATIENTS AND METHODS:

This is a prospective and retrospective study of 23 patients with choledochal cysts who were treated in the gastroenterology and Hepatology teaching hospital in Baghdad in a period extending from April 2000 to November 2010

The patients had been referred from other national health hospital, other hospitals of Iraqi governorate and from private clinics

19 of the cases were studied prospectively and 4 cases were studied retrospectively. 7 cases were males and 16 cases females. The average age at presentation was 37 years (range 4 – 70) years.

All patients were evaluated by history, clinical examination and investigated by the following:

Complete blood picture, ESR, liver function test, renal function test, prothrombin time, partial thromboplastin time, INR and virological assessment.

Radiological examination by chest x-ray, abdominal ultrasound done for all the patients and many of them investigated by MRI / MRCP. CT-scan was done for some of the patients. In patients with suspicion of obstruction of biliary passages based on clinical, biochemical or radiological findings, preoperative ERCP or PTC were performed. Oesphagogastrodudenoscope and EUS were done for some of the patients.

Choledochal cysts were classified according to Todani's classification system (1).

The patients with the cysts were treated either endoscopically or surgically. The clinical presentation, the type of the cysts, type of surgery used the histopathologic features of the cysts, the morbidity and mortality also has been studied.

Follow up were performed by clinic and hospital visits mainly by laboratory investigation with or without ultrasonography.

RESULTS:

There were 23 patients diagnosed with choledochal cysts in the study period. The total admission to the hospital in the same period was 20237 patients and the number of patients with benign biliary diseases was 3659 patients. The patients with choledochal cysts represent 0.1% from the total admission to the hospital and 0.6% from the number of benign biliary diseases.

Age and sex:

There were sixteen female patients (69.5%) and seven male patients (30.4%). The mean age was 25.82 years (range 4-70 years). (See table 1)

Table 1: Age and sex distribution.

Age	Male	Female	Total
1-9 years	2 (8.6 %)	2 (8.6 %)	4 (17.3 %)
10 – 19	2 (8.6 %)	4 (17.3 %)	6 (26 %)
20 – 29	1 (4.3 %)	2 (8.6 %)	3 (13 %)
30 – 39	1 (4.3 %)	2 (8.6 %)	3 (13 %)
40 – 49	-	4 (17.3 %)	4 (17.3 %)
50 – 59	1 (4.3 %)	1 (4.3 %)	2 (8.6 %)
Over 60 y		1 (4.3 %)	1 (4.3 %)
Total	7 (30.4 %)	16 (69.5 %)	23 (100 %)

Distribution of patient's residency

The distributions of patient's residency are as follow:

Table 2: Distribution of residency.

Governorates	Number of patients	Percentage
Baghdad	13	56.5 %
Alsulaymaniyha	2	8.6 %
Diyala	2	8.6 %
Alanbar	2	8.6 %
Almuthanah	1	4.3 %
Karbalah	1	4.3 %
Kut	1	4.3 %
Basrah	1	4.3 %

Presenting symptoms:

Sixteen patients present with epigastric or right hypochondrium pain, fifteen with jaundice, six with fever and one patient presented with epigastric pain, jaundice and abdominal mass. (See table 3)

Table 3: Presenting symptoms of the patients.

Presenting symptoms	Number of patients	Percentage
Epigastric or right hypochondrium pain	16	69.2 %
Jaundice	15	62.2 %
Fever	6	26 %
Epigastric pain with jaundice and abdominal mass	1	4.3 %

Duration of symptoms:

The shortest duration of symptoms was 3 days and the longest was 30 years. The numbers of patients with duration of symptoms of more than 1 year were 9 patients (39.1 %) and those with duration of symptoms less than 1 year were 14 patients (60.9 %).

Diagnostic procedure:

Fifteen patients had hyperbilirubinemia and increased alkaline phosphatase at presentation. Serum amylase, were not systematically performed.

Ultrasound examination done for all patient. MRI/MRCP (12 [52 %]), CT-scan (3 [13 %]), ERCP (11 [47.8 %]), EUS (2 [8.6 %]), OGD (2 [8.6 %]) and PTC (1 [4.3 %]) (see table 4). ERCP was used less frequently in younger than in older patients, 3 of 11 [27.2%] patients who underwent ERCP were below age of 16 years.

Table 4: Diagnostic procedure .

Diagnostic procedure	Number of patient
US	23 (100 %)
MRI / MRCP	12 (52 %)
CT – Scan	3 (13 %)
ERCP	11 (47.8 %)
EUS	2 (8.6 %)
OGD	2 (8.6 %)
PTC	1 (4.3 %)

According to Todani classification, eighteen patients had Type I choledochal cyst, two had Type II, one had Type IV A and two had Caroli's (Type V) disease.(see table 5)

Table 5: Type of choledochal cysts

Type	Number of patient	Percentage
Type I	18	78.2 %
Type II	2	8.6 %
Type III	-	-
Type IV – A	1	4.3
Type V	2	8.6 %

Treatment:

The number of patients underwent surgery are sixteen. Twelve patients underwent Roux-en-Y hepaticojejunostomy with complete excision of the cyst, two patient underwent side to side cystoduodenostomy without excision of the cyst.

One patient underwent Roux-en-Y hepaticojejunostomy as a revision surgery to refashion a previous Roux-en-Y choledochojejunostomy done for her ten months previously in our hospital and one patient underwent Roux-en-Y hepaticojejunostomy as a revision surgery to refashion a previous Loop cystojejunostomy done for her in Basrah 2 years previously. One patient refused surgery.

ERCP and sphincterotomy done for 3 patients with choledochal cyst and associated cystolithiasis and for one patient with cholangitis (70 year old female) who is a poor surgical candidate.

1. Regarding the two patients with Caroli's disease, an ERCP and sphincterotomy done for one of them and PTCD(Percutaneous transhepatic cholangiodrainage) done for the other. (See table 6)

Table 6: Treatment of patients with choledochal cyst.

Treatment	Number of patients
Roux-en-Y hepaticojejunostomy	14 (2 reoperation)
Cystoduodenostomy	2
ERCP and sphincterotomy	5
PTCD	1
Conservative treatment	1

The Roux-en-Y hepaticojejunostomy involve resection of the cyst down to the distal limit of the cyst to ensure that no cyst wall was left behind. The distal common bile duct (CBD) stump was then closed with interrupted sutures, and a Roux-en-Y, end-to-side hepaticojejunostomy was fashioned with single layer of interrupted 3/0 vicyl sutures with extramucosal sutures on the jejunal side. Cholecystectomy was also performed concomitantly. A suction drain was routinely placed in the subhepatic place, and remained in place for three to five days which was removed before patients discharge.

Patient who underwent surgery suffered no major early complications. However, One patient who had a Roux-en-Y choledochojejunostomy 10 month previously presented with obstructive jaundice due to anastomotic stenosis so revision surgery done for her. There was no operative mortality in this study.

The histological examination of the resected cysts revealed cholangiocarcinoma in 2 patients [8.6 %], fibrotic cyst wall in 3 patients and features of acute or chronic inflammation in the remaining patients.

During the period of follow-up, one patient with cholangiocarcinoma died one year after operation. Most of the remaining patients have been regularly reviewed and remaining symptom free.

DISCUSSION:

A Choledochal cyst is a rare disease of the biliary tree. They account for approximately 1% of all benign biliary disease (11). In our study the choledochal cysts account for 0.6% of all benign biliary diseases admitted to our hospital in the study period and most of the patients came from Baghdad, so the general incidence of this disease may be more than this figure because of the presence of other centers in Baghdad and other Iraqi governorates dealing with this disease. Choledochal cysts reported to be more common in Asian populations and in females [1, 4]. Most series show that the most common cyst is the Todani type I (1,4), which is also found in our study . (see table 7). Similarly, we have a preponderance of female patients in our study.

Table 7: Relative incidence of choledochal cysts in our study Compared with Todani et al. [1] study.

Type	Our study	Todani et al.[1] study
I	78.2 %	40 – 85 %
II	8.6 %	2.3 %
III	-	1.4 – 5.5 %
IV	4.3 %	18 – 20 %
V	8.6 %	Rare

In this study, children (≤ 16 years) with choledochal cysts usually present with chronic and intermittent abdominal pain or intermittent jaundice and recurrent cholangitis. Initial presentation in adult (≥ 16 years) is rare . They usually present with non-specific upper abdominal pain associated with obstructive jaundice or cholangitis . Classical triad of abdominal pain, jaundice, and abdominal mass has proved to be rare [(4).] This was confirmed in our study. The main presenting symptoms of our patients were abdominal pain and jaundice as in other studies (Anazawa et al. [(13)], Klotz et al. [(14)], Alonso – Lej et al. [(2)]). (See table 8)

Table 8: Symptoms of our patients compared with others studies.

Symptoms	In our study 23 cases	Anazawa et al[13] 92 cases	Klotz et al. [14] 101 cases	Alonso-Lej et al. [2] 94 cases
Epigastric or RHC pain	69.2 %	43.5 %	41.6 %	64.9 %
Fever	26 %	19.6 %	-	34.0 %
Jaundice	65.2 %	64.1 %	59.4 %	73.4 %

In adult patients with choledochal cyst, there are associated biliary problems such as the presence of cholangiocarcinoma, cystolithiasis, cholangitis and liver cirrhosis with portal hypertension. Indeed, studies in adult have shown that nearly 80% of them present with one of these conditions [(12,15)]. In this study, two patients had cholangiocarcinoma, two patients had liver cirrhosis and four patients had cholangitis (two of them had associated cystolithiasis). The wide variety of presentation among adults is probably secondary to the presence of complications of the cyst, such as stones, cholangiocarcinoma and cirrhosis. This was seen in 61.5 % of our adult patients. (See table 9)

Table 9: Associated conditions in adult patient with Choledochal Cysts in our study compared with other studies.

Associated condition	Our study n = 13	Liu cl et al. [(12)] n = 30	Zhenget al. [(15)] n = 72
Cystolithiasis	2 (15.3 %)	0	12 (16.6 %)
Acute cholangitis	4 (30.7 %)	8 (26.6 %)	29 (40.2 %)
Acute pancreatitis	0	4 (13.3 %)	0
Chronic pancreatitis	0	0	5 (6.9 %)
Liver cirrhosis	2 (15.3 %)	4 (13.3 %)	1 (1.3 %)
Cholecystitis	0	0	54 (75 %)
Cholangiocarcinoma	2 (15.3 %)	9 (30 %)	5 (6.9 %)

Ultrasonography is the best initial method of evaluating dilatation of the intra and extra hepatic bile ducts [(16)]. MRCP has the advantage of being non-invasive and is able to visualize the biliary tree and pancreatic duct as well as ERCP (but without the complications of ERCP). It is hence widely held as the imaging modality of choice for choledochal cyst. However, ERCP was done for most of our patients for its therapeutic role. Our patients commonly present with right hypochondrial pain associated with an obstructive picture of liver function tests, raising the suspicion of choledocholithias which may be dealt with by ERCP.

It is necessary to classify the type of cyst and to recognize the presence of an anomalous pancreaticobiliary duct junction(APBJ), visualization of both the biliary tree and pancreatic duct. For this purpose, direct cholangiography, especially ERCP, is beneficial. Radiographic visualization of both the biliary tree and pancreatic duct prior to surgery is helpful for the surgical manipulation and complete excision of the cyst. In our study a APBJ could not be identified in both MRCP and ERCP. Therefore, no conclusion can be justified concerning the presence or absence of APBJ. While study done by song et al[17] report 27 patients with APBJ out of 5037 underwent ERCP

In this study, CT-scan was done for patients with intra hepatic cysts and for patient with suspicion of malignancy. Regarding the two patients with Caroli's disease, OGD was done for both of them to exclude esophageal varices and PTC was done for one of them for diagnostic and therapeutic (external drainage) role. EUS is not used as standard diagnostic tool for choledochal cysts and in this study were mainly performed during workup of the patient when the diagnosis was still unclear.

Cholangiocarcinoma in choledochal cyst has been reported in 2.5 -28% of patients. However, the overall finding of 8.6% (non intrahepatic) cholangiocarcinoma in our study is comparable with other series. (Table 10) [4, 18, 19, 20]

Table 10: Ncidence of malignancy in choledochal cysts Reported In literature.

Study	No. of patients	No. of patients with malignancies%	Malignancies after internal drainage (% of all malignancies)	Age at presentation of malignancy
Our study	23	2(8.6)	0 (0)	42(35-50)
Lipsett et al.[3]	42	3 (10)	0 (0)	Adult
Todani et al.	82	8 (10)	3 (38)	-
Janet al[17]	80	8 (10)	3 (38)	50 (32-81)
Bismuth and Krissat[19]	48	6 (13)	2 (23)	39 (17-57)

The concept of treatment of extrahepatic choledochal cysts has changed in the past 30 years because of a persistent high risk of malignancy after drainage procedure [17,20]. In view of the high risk of cholangiocarcinoma, the state of the art treatment of extrahepatic choledochal cysts is primary excision with construction of a biliary digestive anastomosis[17,20].Fourteen of our patients have had resection of their cysts with a Roux-en-Y hepaticojejunostomy. A wide – caliber hepaticojejunostomy must be done to the normal bile duct using a meticulous surgical technique to avoid leaks. All this will ensure a patent biliary bypass. All these patients are currently doing well. Although, the remaining patients who underwent cystoduodenostomy or ERCP and sphincterotomy became well and symptom free after these procedure, excision of extrahepatic cysts after internal drainage (cystoduodenostomy, ERCP and sphincterotomy) is recommended, even in the absence of symptoms[17,20].

CONCLUSION:

Choledochal cyst is rare disease. The main presenting symptoms are abdominal pain and jaundice. Although the initial presentation in adults is rare, it does occur. The presentations are variable and are due to presence of associated diseases. In view of the high risk of cholangiocarcinoma, early resection and not internal drainage is the appropriate treatment of type I and II and extrahepatic part of type IV biliary cyst. The correct surgical option would be total excision of the cyst and reconstruction with a Roux-en-Y hepaticojejunostomy

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