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Management of choledochal cysts in gastroenterology and hepatology teaching hospital

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ABSTRACT

Background: Choledochal cysts (CCs) are uncommon cystic dilatations of extra and/or intrahepatic biliary tree which may occur singly or in multiples throughout the bile ducts.The incidence of choledochal cyst is 1 in 100,000150,000 live births in the western population, but reported to be as high as 1 in 13,500 live births in the United States and 1 in 15,000 in Australia.The incidence is higher in Asian population with an incidence of 1 in 1000, of which about two-third cases are reported from Japan. CCs are usually diagnosed in childhood and about 25% are detected in adult life.**Aim of study**:To analyze the experience of managementof choledochal cysts disease in gastroenterology and hepatology teaching hospital in BaghdadMedical City from January 2003 to March 2015.**Materials and methods:**A descriptive prospective and retrospective study of (40) patients with choledochal cysts disease who were admitted and treated in the gastroenterology and hepatology teaching hospital in Baghdad Medical City from January 2003 to March 2015.The retrospective study period started from January 2003 to November 2011, while the prospective study started from December 2011 to March 2015. Twenty-three cases studied prospectively, while 17 cases studied retrospectively.**Results:** A forty cases were studied, 28 (70 %) were females and 12 (30 %) were males, about the age distribution, the patients age range from (1- 60),the mean age was (17.26) about the residency they were mainly from Baghdad in 16 cases (40%), the most common presenting symptoms areepigastric pain 26 patients (65 %) and jaundice in 20 patients (50 %).The U ? S and MRI ? MRCP are the most common imaging used. According to the Todani classification of CCs disease we found the type I CCs is the most common type of the cyst (77.5 %).Total excision of the cyst wall with a Roux-en-Y hepaticojejunostomy (RYHJ) is the treatment of choice for choledochal cysts.**Conclusions:** The Clinical suspicion of CCs should be followed by early diagnosis and management in view of life-threatening complications and high risk of malignancy. MRCP is the imaging modality of choice except in choledochoceles, which needs multiple imaging modalities before diagnosis. A complete excision of the extrahepatic system and RYHJ is the treatment of choice in type I and most of type IV Ccs.

Key Words: Etiopathogenesis, choledochal cysts, classification, excision,Roux-en-Y hepaticojejunostomy

Introduction:

Definition: Choledochal cysts (CCs) are uncommon cystic dilatations of extra and/or intrahepatic biliary tree which may occur singly or in multiples throughout the bile ducts.[1]**Incidence**:The incidence of choledochal cyst is 1 in 100,000150,000 live births in the western population, but reported to be as high as 1 in 13,500 live births in the United States and 1 in 15,000 in Australia.The incidence is higher in Asian population with an incidence of 1 in 1000, of which about two-third cases are reported from Japan.[2]**CLASSIFICATION:**Initial classification by Alonso-Lej et al. in 1959 described 3 types of CCs. Later Todani et al. in 1977 modified it by adding type IV and V.Modified Todani et al. classification is most commonly used by surgeons.[3]

1.Type I CCs:are fusiform dilatations of extrahepatic biliary tree, and is the most common Type of CCs comprising about 50%-80% and further sub classified into 3 types.

- Type IA: is cystic dilation of entire extrahepatic biliary tree with sparing of intrahepatic ducts. Cystic duct and gall bladder arises from the dilated common bile duct (CBD).
- Type IB: is focal, segmental dilation of extrahepatic biliary tree.
- Type IC: is fusiform dilation of entire extrahepatic biliary tree extending into intrahepatic duct.

2. Type II CCs:are saccular diverticulumof the CBD.It's a rare type of CC consisting about 2%.

3. Type III CCs: also termed choledochoceles, comprising about 1.4%-4.5%, represents cystic dilation of intramural portion of distalCBD with bulge into the duodenum.

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4. Type IV CCs:occur in about 15%-35% and are further subclassified into type IVA andtype IVB. Type IVA is the second most common CCs andis described by both intrahepatic and extrahepatic dilation of biliary ducts. Type IVB represents multiple dilation of extrahepatic biliary tree only.

5.Type V CCs: known as Caroli's disease represents multiple dilation of intrahepatic biliary ducts consisting 20%. It is termed Caroli's syndrome when associated withcongenital hepatic fibrosis, which then may present with cirrhosis and its manifestations. Lilly et al. described an entity called "forme fruste" CCs,(an incomplete or atypical form of a disease or a disease that is spontaneously arrested before it has run its usual course) where the patients present with typical symptoms of CCs and are associated with abnormal pancreaticobiliary ductjunction (APBDJ) but without dilation of biliary ducts.[3]

Etiopathogenesis: Etiology of CCs is an ongoing debate with both congenitaland acquired theory supporters. The most commonlyproposed theory is Babbitt's theory, where CCs are supposedto be caused by anAPBDJ in which the pancreatic duct joinsthe bile duct (1-2 cm) proximal to the sphincter of oddi. This long common channel allowspancreatic juice reflux into biliary system and cause increasedpressure within the common bile duct(CBD) resulting in ductal dilation. But this theory is questioned by some authors because APBDJ is observedin only (50-80%) cases of CCs. Obstruction ofdistal CBD is another theory, which is supported by studieson animal models.Sphincter of oddi dysfunction reportedin some studies may predispose to Ccs.**clinical Presentation:** CCs most commonly present in childhood and about 25%patients present in adulthood. The classic triad of symptoms,which includes pain abdomen, palpable abdominal mass, andjaundice, is seen in less than 20% of cases. An 85% of childrenhave at least 2 features of classic triad, whereas only 25% ofadults present with at least 2 features of the classic triad.[5]**Malignancy:** The increased risk of malignancy in CCs is well known. Thereported incidence varies from 2.5% to 17.5% in patients withCCs.The incidence of malignancy increases with age,supposed to be 0.7% in the first decade of life to 14.3% after20 years of age, which means early diagnosis and treatmenthas a favorable outcome.Todani et al. observed 68% of malignancyin type I, 5% in type II, 1.6% in type III, 21% in type IV, and6% in type V CCs. Malignancy occurs in 1239% of "FormeFruste" patients. Malignancy in Caroli'sdisease is reported to be about 7-15% and in choledochocelesabout 2.5%.[6,7]

Diagnosis: Abdominal ultrasound (US) scan is the first step towardconfirmation of diagnosis. Sensitivity of US is about 71-97%. It is also the preferred investigation in postoperativesurveillance. Computed tomography (CT) is highly accurate and alsohelp in planning surgical approaches. It delineates well theintrahepatic biliary dilation in type IVA and Caroli's diseaseand also the extent of intrahepatic dilation, which helps insurgical planning, such as segmental lobectomy, in case oflocalized intrahepatic biliary ductal dilation. Endoscopic retrograde cholangiopancreatography (ERCP)is reported to be the most sensitive diagnostic modalityfor CCs.But the sensitivity decreases in case ofrecurrent inflammation and scarring where the procedurebecomes difficult. ERCP in CCsalso need large amount of dye to fill cyst, which increases thechance of cholangitis and pancreatitis.MRCP is regarded as the "goldstandard" for the diagnosis of CCs.Sensitivity hasbeen reported to be as high as 90-100 %.Type III CCs may need multiple modalities beforemaking a diagnosis. Upper gastrointestinal series mayshow a filling defect due to bulge into duodenal lumen.Endoscopy and ERCP demonstrates bulging and also dilatedintramural CBD.[8]

Management: Complete excision of the cyst and RYHJ is now consideredthe surgery of choice in most of the CCs. The surgical approach in type IVA is still debatable. Visser et al. suggested excision of extrahepatic component only with hepatico jejunostomy in case of type IVA CCs irrespective of the changes.However, in case of extensive intrahepatic dilation with complications, such as stones, cholangitis, or biliary cirrhosis, other options, such as hepatic resection in case of unilobar disease and liver transplantation in bilobar disease should be considered.Nowa days, cyst excision and RYHJ are also done laparoscopically. Type II CCs are managed by simple excision. Usuallythese cysts are ligated at the neck and excised withoutthe need for bile duct reconstruction. Type III CCs were historically treated by transduodenal excision and sphincteroplasty. But recently endoscopic sphincterotomy is accepted to be sufficient treatment but patient should be under endoscopic surveillance since malignancy has been reported in choledochoceles. Ohtsuka et al. observed malignancy in 3 of 11 patients with choledochoceles. In Caroli's disease, when the intrahepatic duct dilation is localized and without congenital hepatic fibrosis, segmental hepatectomy can be done.[9,10]

Aim of study: To analyze the experience of managementof choledochal cysts disease in gastroenterology and hepatology teaching hospital in Baghdad Medical City from January 2003 to March 2015.

Patients and methods: A descriptive prospective and retrospective study of (40) patients with

choledochal cysts disease who were admitted and treated in the gastroenterology and hepatology teaching hospital (GHTH) in Baghdad Medical City from January 2003 to March 2015. The retrospective study period started from January 2003 to November 2011, while the prospective study started from December 2011 to March 2015.In our study (23) cases were studied prospectively and (17) cases were studied retrospectively.Twelve cases were males (30%) and 28 cases were females (70%). The average age at presentation were (17.26) years, (range 1-60) years. In this study all cases were evaluated by history, clinical examination (as epigastric pain, jaundice, fever, abdominal mass and others) and different investigations including:Complete blood picture, liver function test, renal function test, serum amylase, coagulation profiles, virology and others.Radiological imaging including abdominal ultrasound done for all the patients, and half of them investigated by MRI/MRCP. CT-scan done in twelve patients. Other imaging like EUS, OGD, ERCP and PTC are used according to the patients need. Choledochal cysts were classified according to the Todani classification system.In most cases the operative finding was type I CC which treated by total cyst excision from common hepatic duct down to the retropancreatic portion of the common bile duct with RYHJ and cholecystectomy. Results:Forty patients diagnosed with choledochal cysts disease in the study period in GHTH in BaghdadMedical City from January 2003 to March 2015.The number of patients with benign biliary diseases admitted to our hospital in the same period were (4691) patients so, the percentage of the patients with CC disease represent about (0.8%) from the number of benign biliary diseases.

Age and sex distribution:

There were (28) females (70 %) and (12) males patients (30%). The age range from 1 year to more than 60 years, (the mean age was 17.26 years). (See table 1)

Table 1. Age and sex distribution

Age (year)	Male	Female	Total
1-10	5 (12.5 %)	11 (27.5%)	16 (40%)
11-20	3 (7.5 %)	5 (12.5 %)	8 (20 %)
21-30	1 (2.5 %)	2 (5 %)	3 (7.5 %)
31-40	1 (2.5 %)	4 (10 %)	5 (12.5 %)
41-50	1 (2.5 %)	3 (7.5 %)	4 (10 %)
51-60	1 (2.5 %)	2 (5%)	3 (7.5%)
> 60	-------	1 (2.5 %)	1 (2.5 %)
Total	12 (30 %)	28 (70 %)	40 (100 %)

Distribution of patient's residency: The distributions of patient's residency according to the Iraqi governorates are listed in figure 1.

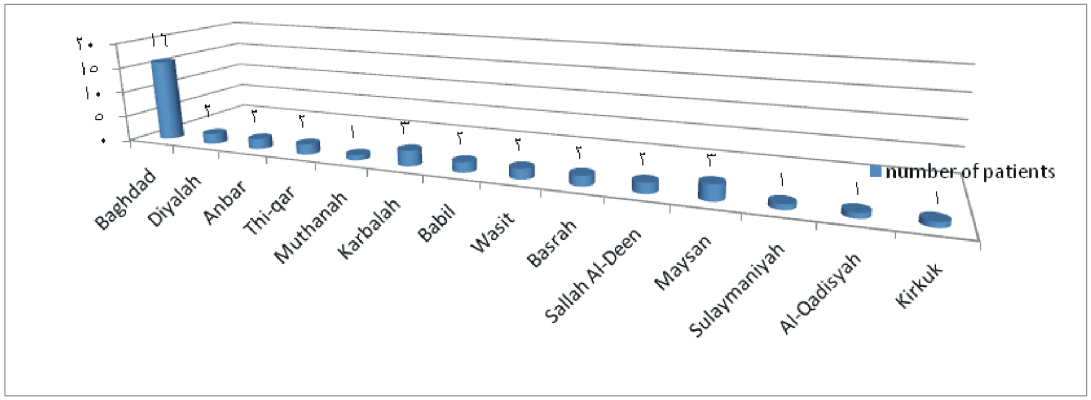

Figure (1).Distribution of patient's residency

Presenting symptoms and signs: The most common presenting symptoms of CCs in present study are epigastric or right hypochondrial pain, jaundice and fever. (See table 2)

Table 2.Presenting symptoms and signs of the patients

Presenting symptoms and signs	Number of patient	percentage
Epigastric or right hypochondrial pain	26	65 %
Jaundice	20	50 %
Fever	8	20 %
Epigastric pain and Jaundice	10	25 %
Epigastric pain, jaundiceand abdominal mass	2	5 %

In our study we found that some patients may present with more than one or two symptoms and signs at the same time, while the other patients present with single symptom or sign.

The duration of symptoms:In our study the shortest duration of symptoms was 10 days and the longest duration was 25 years.The number of patients with duration of symptoms less than 1 year were (24) patients (60 %).

Complications and associated conditions inpatients withcholedochal cysts:

In present study we found that 30 patients (75%) associated with different biliary problems.(See figure3)

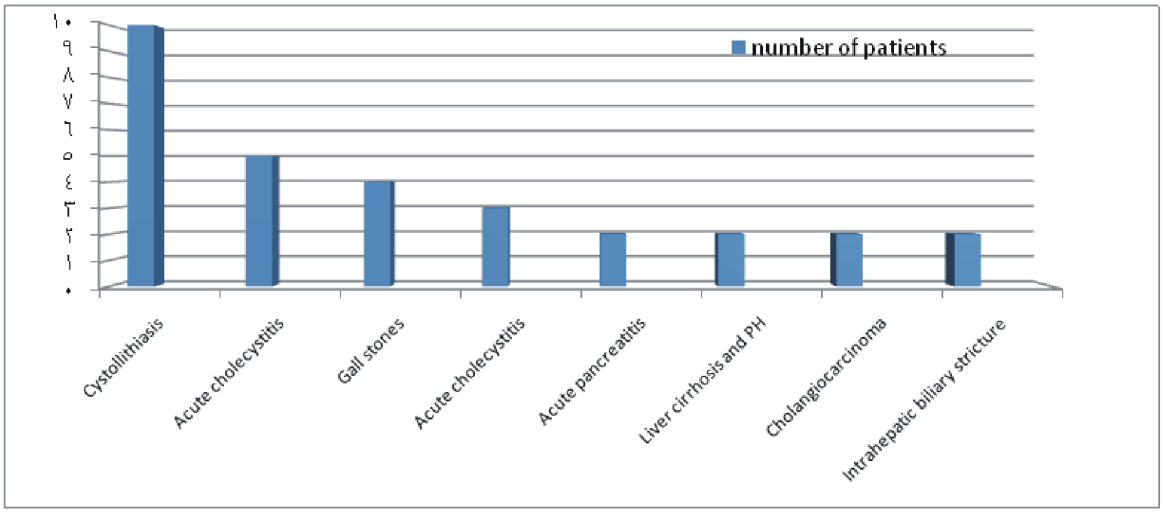

Figure (2). Complications and associated conditions of patients with CCs

In this study we found one child with CCs disease (Caroli's disease) associated with portal hypertension (with esophageal varices) and medullary spongy kidney. Age of child was 9 years.

Diagnostic procedures: Thirtypatients presented with hyperbilirubinaemia and increased alkaline phosphatase at presentation after doing the specific and general biochemical laboratory investigations for all our patients. Serum amylase level was performed in 10 patients only and the results were within normal limits.An ultrasound examination was done for all the patients (100 %), while theMRI/ MRCP done for half of the patients (50 %), and theCT-scan done for 12 of 40 patients (30 %). (See table 3)

Table 3.Diagnostic procedures.

Diagnostic procedures	Number of patients	Percentage
US	40	100 %
MRI/MRCP	20	50 %
CT- Scan	12	30 %
ERCP	5	12.5 %
EUS	5	12.5 %
OGD	3	7.5 %
PTC	1	2.5 %

Types of choledochal cyst:

In this study we depend on the Todani classification, so we found about thirty one patients (77.5 %) had Type I choledochal cyst (the most common type).(Seefigure 4)

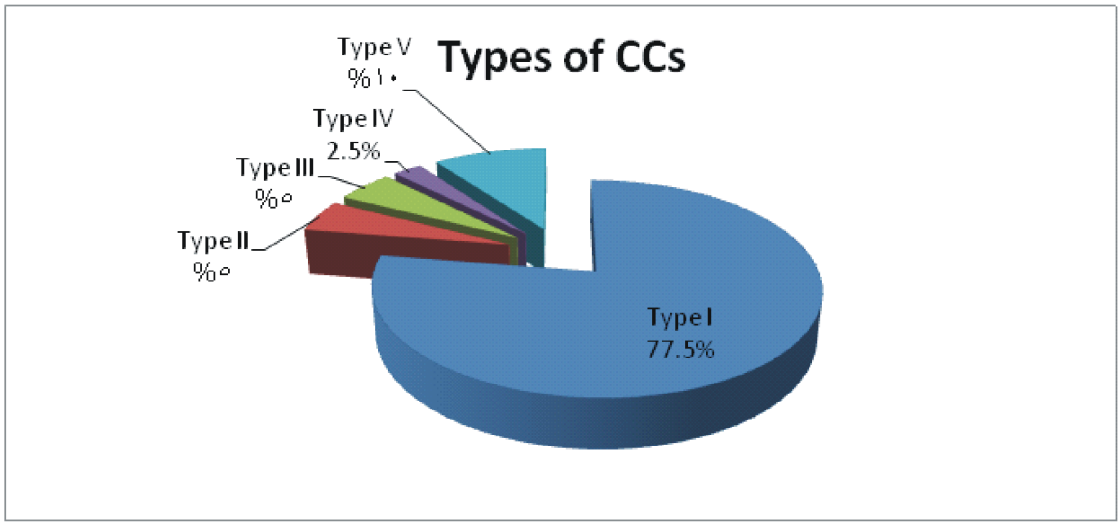

Figure (3). Types of choledochal cysts

Treatment:

Regarding the treatment of choledochal cysts we found about thirty-four patients (85 %) treated by surgical intervention, of those patients,thirty-one of them underwent total cyst excision from the common hepatic confluence down to the retropancreatic portion of the common bile duct withRYHJ,two of themunderwent side to side Cystoduodenostomy, without excision of the cyst, and one patient with type III CC treated surgically by anterior duodenotomy with excision of the cyst and transduodenal sphincteroplasty with primary duodenal closure. Of those thirty-onepatients with CCs who treated surgically, two patients underwent RYHJas arevisional procedure to a previous RYHJ done forthem years agodue to development of postoperative biliary stricture and subsequent cholangitis (one of them the surgery done in our hospital before 2 years ago, and the other patient in Kirkuk hospital before one year ago). Another patient underwentRYHJ as a revisional surgery to refashion a previous loop cystojejunostomydone for him in Basrah before 3 years ago(also developed postoperative stricture).In patient with type I CC with extension of the dilatation to the extrahepatic portion of the right and left hepatic ducts (HD) so, total excision of the cyst done distally down to the retropancreatic portion of the CBD and proximally up to the hepatic hilum with separated right and left hepatic duct RYHJ. In two cases with Ccs disease there is associated intrahepatic biliary stricturefor which the CCswastotally excised with ductoplasty done at the hepatic confluence with slight extension to the left hepatic duct in order to facilitate the reconstruction with RYHJ.Also ductoplasty done in other patient with CC, in this case the hepatic confluence occur between the left HD and the right anterior duct, and the right posterior duct joined separately to the CHD, so the bilateral RYHJ done to both right posterior duct and to the common hepatic confluence after ductoplasty of the common hepatic septum.In this study one female patient with age of 37 years refuse surgery and treated conservatively.ERCP and sphincterotomy done for:Two patients with choledochal cystsand associated cystolithiasis,one patient with type IIICC,one patient with cholangitis (50 years old female) who was a poor surgical candidate, and for one patient with Caroli's disease.Regarding the four patients with Carole's disease, anERCP and sphincterotomy done for one of them,and PTC done for the other (due to the CBD stones).Follow up done for the other two patients with Caroli's disease without any surgical intervention. (See table 4).

Table 4.Treatment of patients with choledochal cysts disease

treatment	Number of patients	percentage %
Cyst excision with RYHJ	31	77.5%
Cystoduodenostomy	2	5%
Transduodenal cyst excision with sphincteroplasty	1	2.5 %
ERCP and sphincterotomy	5	12.5 %
PTC	1	2.5 %
Conservative treatment	3	7.5 %

Three of five patients who underwent ERCP (two patients with CCs and cystolithiasis and the other with cholangitis) also finally treated with RYHJ.In this study all the specimens were send for histopathological examinations and the results were normal except two patients had associated cholangiocarcinoma with CCs disease, and treated by total excision of the extrabepatic biliary system including the cyst and the tumor,with RYHJ .***Postoperative complications:***In this study no major and life threatening postoperative complications were developed. Nineteen patients (47.5%)were developed early or late complications.The most common early complications are bile leak which occur in 2 patients (5 %) and treated conservatively and improved within 3-5 days, two patients (5 %) presented with ileus who improved after few days on conservative treatment,while the rare complication is the development of Chylous ascites (2.5 %) and this treated conservatively by fat free diet, octreotide and peritoneal dialysis (PD)catheter for one month.Two patients developed acute pancreatitis (5 %), and one patient presented with acute cholangitis (2.5 %) and both these conditions treated conservatively. Also three patients developed chest infection (7.5 %) and other three patients (7.5 %) presented with wound infection (7.5 %). There was no intraoperative or postoperative bleeding.Regarding the late complications, one patient (2.5 %) presented with picture of obstructive jaundice due to stricture at the site of anastomosis after Roux-en-Y hepaticojejunostomy so,a revisional surgery done for himafter two years of first operation with a good results.

Three patients developed incisional hernias (7.5 %), and one of the patients (2.5 %) who treated for CC and associated cholangiocarcinoma developed abdominal distension, ascites and abdominal pain (tumor recurrence) and treated by chemotherapy and paracenthesis. There was no operative mortality in this study, but in one child below the age of 16 year with type III CC who treated by ERCP, died after 3 days of the procedure and the cause of death most likely due to iatrogenic perforation and subsequent septicemia. Discussion:Choledochal cysts is a congenital abnormality of biliary tree that requires surgical intervention to prevent hepatobiliary and pancreatic complications.Choledochal cysts disease account for approximately(1%) of all benign biliary disease.[12]In this study the CCs account for approximately (0.8%) of all benign biliary disease admitted to our hospital in the study period and this incidence may be less than the real incidence, because most of the patients with CCs disease came from Baghdad city to our hospital, so the general incidence of this disease may be more than this figure because of the presence of other centers in Baghdad and also other Iraqi governorates dealing with this disease.Gastroenterology and hepatology teaching hospital is the most common center in Iraq dealing with different biliary tract diseases including CCs disease.The age of patients in this study is ranging from (1-60) years, and the mean age was (17.26) year. The peak incidence of CCs disease was in the first decade of life, (1-10 year). This result was similar to other studies like Anazawa et al. and Klotz et al.

Regarding the sex distribution, we found most patients with CCs disease in our study were females (28 females and 12 males), and this is mimicking the results of other studies like Liu et al.[4]and Todani et al.[1]

The main presenting symptoms in our patients were abdominal pain in (65 %), and jaundice (50%), and this results compatibleto other studies results such as Anazawa et al.[13] Klotz et al.andAlonso-Lej et al.(See table 5)

Table 5. Symptoms and signs of our patients compared with others studies

Symptoms and signs	Our study 40 cases	Anazawa et al. 92 cases	Klotzet al. 101 cases	Alonso-Lejet al. 94 cases
Epigastric or RHC pain	26 (65%)	43.5 %	41.6 %	64.9 %
jaundice	20 (50%)	64.1 %	59.4 %	73.4 %
fever	8 (20%)	19.6 %	23 %	34 %
epigastric pain and jaundice	10 (25%)	28 %	27.6 %	41.4 %
Epigastric pain, jaundice And abdominal mass	2 (5%)	-------	-------	8 %

In our study we found that the shortest duration of symptoms was 10 days as an abdominal pain with development of jaundice and sometimes nausea and/or vomiting, while the longest duration of symptoms about 25 years as development of recurrent attacks of cholangitis with picture of obstructive jaundice and fever,and sometimes pictures of acute cholecystitis due to gall stones formation. These results in our study was similar to the results in other comparative studies such as Lipsett et al. [14]Thenumber of patients with duration of symptoms of more than 1 year was16 patients (40%) and those with duration of symptoms less than 1 year were 24 patients (60 %). This differences in the percentages because the majority of cases of CCs disease occurs in the first decade of life [about 16 patients (40 %)].

Regarding the CCs disease, there are several associated biliary problems such as the presence of a cystolithiasis, gall stones with acute cholecystitis, acute cholangitis, and acute pancreatitis, liver cirrhosis with portal hypertension, cholangiocarcinoma and intrahepatic biliary stricture. In many studies of CCs disease, we shows that nearly 75% of patients presented with one of these conditions, like Liu et al.[2]study and Zheng et al.study.[15]In present study we found that 30 patients of 40 associated with and complicated by one or more of these conditions as in (table 6).According to these results we found that a lot of presentations in patients with CCs are probably secondarily to the presence of these conditions mentioned above. This was seen in (75 %) in our patients.

Table 6.Complications and associated conditions in patients with choledochal cysts in our study compared with other studies

Associated conditions	Our studyN= 30	Liu et al.N= 28 (T.N.=163)	Zheng et al.N= 72
Cystolithiasis	10 (33.3 %)	10 (35.7%)	12 (16.6 %)
Acute cholangitis	5 (16.6 %)	8 (28.5 %)	29 (40.2 %)
Gall stones	4 (13.3 %)	2 (7.1 %)	5 (6.9 %)
Acute cholecystitis	3 (10 %)	0	54 (75 %)
Acute pancreatitis	2 (6.6 %)	4 (14.2%)	0
Liver cirrhosis with PH	2 (6.6 %)	4 (14.2 %)	1 (1.3 %)
cholangiocarcinoma	2 (6.6 %)	9 (32.1 %)	5 (6.9 %)
Intrahepatic biliary Stricture	2 (6.6%)	10 (35.7)	1 (1.3 %)

In this study the laboratory investigations and ultrasonography done for all the patients with CCs disease. The ultrasound examination is considered the best initial method for evaluating the dilatations of the intra and extra hepatic biliary tract, and this was similar to other studies results, like Akhan et al.[16]MRCP is considered the gold standard imagingmodality for diagnosis the CCsbecause it avoid ionizing radiation and is also noninvasive when compared with ERCP with no complications of pancreatitis or cholangitis.The ERCP used for patients complaining from right hypochondrial pain associated with picture of obstructive jaundice on the LFT, and cystolithiasis on abdominal U/S and all these findings raises the suspicion of CBD stones, also the ERCP used for patient with type IIICC disease. In addition to it is use in patient with cholangitis and in other patient with Caroli's disease who developed jaundice (due to CBD stones),and both these conditions occur in our study.In patients with choledochal cysts disease it is necessary to classify the types of the cyst, and to recognize the presence of an anomalous pancreaticobiliary duct junction, i.e. visualization of both the biliary tree and pancreatic duct. For this reason, a direct cholangiography, especially the ERCP, is of benefit. By using the radiological imaging, the visualization of both the biliary tree and pancreatic duct before the Surgical interventions is helpful for both the surgical manipulation and complete cyst excision.[9]In present study the anomalous pancreaticobiliary duct junction could not be identified in both MRCP and ERCP, and this may be due to lack of experience to identifying this anomaly and in our hospital most physicians doing ERC rather than ERCP. Therefore, no conclusion can be justified concerning the presence or absence of an APBJ. The role of CT-scan in our study is to assess the patients with intra hepatic cysts, to exclude other abnormalities of hepatobiliary system and for patients with suspicion of malignancy.Esophagoduodenoscopy was done for two patients with CCs that associated with liver cirrhosis and portal hypertension to exclude esophageal varices and for one child with CCs disease and esophageal varices due to portal hypertension.PTC is used for one of the patients with Caroli's disease for an external drainage due to intra hepatic biliary stones. EUS is used when the diagnosis of CCs is still in doubt, like in cases of CBD dilatation with stones inside or in cases of mass suspicion and for dilated biliary system without obvious cause. The most common cyst type is the Todani type I CCs followed by other cyst types, these results also found in ourstudy, were we have 31 patients (77.5 %) with type I CCs.LikeTodani et al.[1]study (See table 7)

Table 7. Relative incidence of choledochal cysts types in our study compared with Todani et al.study

type	Our study	Todani et al.study
I	31 (77.5 %)	40-85 %
II	2 (5 %)	2.3 %
III	2 (5 %)	1.4-5.5 %
IV	1 (2.5 %)	18-20 %
V	4 (10 %)	Rare

Inpresent study about two patients (5%) with CCs disease developed cholangiocarcinoma, and this percentage is considered relatively low in comparison with percentage of malignancy in other studies results.(See table 8)

Table 8. Incidence of malignancy of choledochal cysts in our study compared with other studies

The study	Total number of patients	Number of Patients with malignancy	Malignanciesafter internal drainage	Age at presentation of malignancy
Our study	40	2 (5 %)	0 (0%)	55 (50-60)
Lipsett et al.	42	3 (10%)	0 (0%)	Adult
Todani et al.	82	8 (10%)	3 (38%)	-------
Janet et al.	80	8 (10%)	3 (38%)	50 (32-81)
Bismuth and Krissat et al.	48	6 (13%)	2 (23%)	39 (17-57)

Regarding the management of CCs disease we foundIn this study, about thirty-one patients (77.5 %) with CCs underwent total cyst excision (from common hepatic duct confluencedown to the retropancreatic portion of CBD) with a RYHJ.[11]Total removal of the CC wall sometimes require a bilateral anastomosis between Roux-en-Y loop of jejunum and right and left hepatic ducts and this occur due to complete separation of the hepatic confluence during total cyst excision. In this studyductoplasty done during surgery to the patients with CCs disease associated with intrahepatic biliary stricture and this will facilitate the secure biliary anastomosis with good results.Also ductoplasty done for the case of CC in which there's biliary anomaly were the right anterior hepatic duct joined the left hepatic duct forming the common hepatic duct (CHD) and the right posterior duct enter the CHD separately so, during surgery the CC wall completely excised and ductoplasty of the CHD with slight extension to the left hepatic duct with a bilateral RYHJ anastomosis done with both, the CHD and right posterior duct.In one case with type III CC a transduodenal sphincteroplasty done with cyst excision through duodenotomy incision. All these patients are currently doing well, except two patients presented with bile leak for several days and respond well to conservative treatment and one patient developed Chylous ascites after surgery and treated conservatively with fat free diet, octreotide and PD catheter for about one month duration. One patient developed jaundice due to anastomotic stricture (2 years after the primary treatment with RYHJ done in our hospital) with good benefit after arevisional procedure.Also we have two patients treated by Cystoduodenostomy due to extension of the cystic dilatation to the intra hepatic biliary tree so, no benefit from total excision of extra hepatic cyst because the risk of cholangiocarcinoma is still present in the dilated part of the intra hepatic cyst, and also because of severe adhesions between the choledochal cysts and the surrounding structures like portal vein and hepatic artery. This also confirmed by Visser et al.[6]

Revisional surgery in form of RYHJ done for three patients in our study with good results and outcome.Five patients treated with ERCP and sphincterotomy (two of them with cystolithiasis, one with type III CC, one with cholangitis and the other with Caroli's disease due to development of the CBD stone formation), and only one patient with Caroli's disease treated by PTC (due to obstructive jaundice and CBD stone formation). In this study one patient refused surgery and treated conservatively together with other two cases of Caroli's disease.(See table 9)

Table 9. Treatment of CCs in our study compared with Sharma et al.[11]

Treatment	Our studyno = 40	Sharma et al study no = 35
Cyst excision with RYHJ	31 (77.5 %)	26 (74.2 %)
Cystoduodenostomy	2 (5 %)	2 (5.7 %)
Choledochoduodenostomy	-------	5 (14.2 %)
Internal drainage (ERCP)	5 (12.5 %)	0
External drainage (PTC)	1 (2.5 %)	2 (5.7 %)
Transduodenal sphincteroplasty with cyst excision	1 (2.5 %)	---------

Although, the patients who underwent Cystoduodenostomy, and an ERCP and sphincterotomy or PTC became well and symptom free after these procedures, but the total excision of extra hepatic cystis recommended and mandatory, even in absence of symptoms in order to avoid the risk of malignancy.All patients are treated and followed properly after surgery and most of them discharged well after 6-7 days, except some patients delayed for several days due to development of complications.[16]

In present study there weresome patients developed early and latepost-operative complications of CCs surgery and all the early complications were treated successfully in the same admission with good results, except the Chylous ascites which treated conservatively for about one month. Regarding the late complications, one patient developed pictures of obstructive jaundice due to stricture at the site of anastomosis. Another patient developed intestinal obstruction and three patients presented later with incisional hernia.As mention there was no operative mortality during surgical intervention for CCs disease in present study, only one child female with age of 3 yearsdied because of ERCP complications (perforation and sepsis).[17]

Conclusions:

The Clinical suspicion of CCs should be followed by early diagnosis and management in view of life-threatening complications and high risk of malignancy. Thedelay in diagnosis will worsen the prognosis.Abnormal pancreatobiliary duct junction, distal CBD obstruction, and sphincter of oddi dysfunction are proposed to be the etiologic factors. MRCP is the imaging modality of choice except in choledochoceles, which needs multiple imaging modalities before diagnosis.A complete excision of the extrahepatic system and RYHJ is the treatment of choice in type I and most of type IV CCs. Internal or external drainage of cysts should be considered only in case of emergency and as a palliative procedure. Patients who had previously undergone cystenterostomy should undergo reoperation for complete cyst excision and RYHJ. Type II cysts need simple cyst excision, whereascholedochoceles are managed by endoscopic sphincterotomy with a follow-up endoscopic surveillance. Few cases of localized intrahepatic type IVA CCs and Caroli's disease with complications should be considered for hepatic resection. Diffuse intrahepatic disease with complications in type IVA and Caroli's disease should be offered liver transplantation.

Recommendations :

1. Choledochal cysts are uncommon, but when encountered, they may appear nonspecifically rather than classically. A high index of suspicion will avoid a delay in diagnosis.
2. The imaging modality of choice for diagnosing and characterizing choledochal cysts is magnetic resonance cholangiopancreatography (MRCP).
3. The number of patients diagnosed with CCs might be raised if there was increased interest in the diagnostic criteria for APBJ.
4. Delayed diagnosis may have a variety of undesirable sequelae, including biliary cirrhosis, cholangiocarcinoma, pancreatitis, and cholangitis.
5. To avoid these complications, choledochal cysts should be treated by complete excision, whenever possible, with reconstruction using internal drainage.
6. Postoperative follow-up is essential to detect development of any complications, such as cholangitis, anastomotic stricture, or intrahepatic choledocholithiasis.

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