Original Paper

Experiences in the Management of Choledochal Cyst in Pediatric Patients in a Tertiary Care Center of Bangladesh Armed Forces

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Abstract

Introduction: Choledochal cyst (CC) is a rare congenital abnormality of the biliary tract, for which there is no standardized surgical treatment. If left untreated it may cause bile duct (BD) obstruction to cholangiocarcinoma. For the commonest type of Choledochal cyst (Type I and IV) the treatment is surgery, although, the reconstruction technique of the BD has no gold standard.

Objective: To assess the outcome of surgical treatment of choledochal cyst in pediatric cases in 8 years period in CMH Dhaka, a tertiary referral hospital of Bangladesh Armed Forces.

Materials and Methods: This cross sectional study was carried out in the department of Paediatric Surgery at CMH Dhaka on 20 children of under 10 years having choledocal cysts. The study period was from March 2006 to February 2014.

Results: Total of twenty patients aged 3 years to 10 years was included in the study. Out of them 14(70%) were females and 6(30%) were males. Fifteen (75%) patients presented with recurrent abdominal pain, 14(70%) with recurrent jaundice, 4(20%) patients with abdominal mass as initial symptoms. Twelve (60%) patients were diagnosed by Ultrasonogram (USG), rests were by Contrast Enhanced Computed Tomography (CECT) and Magnetic Resonance Cholangiopancreatography (MRCP). Eighteen patients presented with type 1 choledochal cyst and 2 of them had type IV choledochal cyst. All the patients were submitted to cyst excision and Roux-en-Y hepaticojejunostomy. Among them one patient required refashioning of anastomotic site to treat bile leakage.

Conclusion: Early diagnosis and referral is essential to prevent complications and grave consequences and prognosis of these cases after surgery is good.

Key-words: Choledochal cyst (CC), Bile duct (BD), Cholangiocarcinoma.

Introduction

Choledochal cyst (CC) is a rare congenital abnormality of the biliary tract and it accounts for approximately 1% of all benign biliary diseases[1]. It is defined as an aneurysmal dilatation of the biliary tree involving either extrahepatic or intrahepatic biliary ducts or both[2,3]. It's prevalence, although as high as 1:1000 in Asian populations, is only 1:100000 to 1:150000 in western populations[3]. As the condition is not confined to the extrahepatic bile duct, the term “choledochal cyst” is in fact a misnomer and ‘biliary cyst’ is probably more appropriate. Most of the reported cases in the world come from East Asia. It is common in Japan[4] with an incidence of 1:1000. The first reported case of choledochal cyst was described in 1723 by Vater & Ezler[5]. Initial classification by Alanso-Lej et al[6] classified choledochal cysts into three main types, type I–III on clinical and anatomic findings in 1959. This classification was further updated by Todani et al in 1977 who described five main types by adding type IV and V with several subtypes according to location of biliary duct dilation[7]. Type I is further sub classified into A, B, C, but does not influence surgical management and outcome. Todani classification does not include lesion Type VI which is an isolated cyst of the cystic duct, an extremely rare lesion. Choledochal cyst is a rare congenital anomaly of the bile duct[1] for which there is no standardized surgical treatment[2]. If left untreated CC has complications that range from BD obstruction to cholangiocarcinoma with a M:F 1:3–4, female predominance, 75% of which are diagnosed in childhood[1,3].

The treatment of the CC will vary accordingly to the Todani's classification[4]; for the commonest type of Choledochal cyst (Type I and IV) the treatment is surgery, although, the reconstruction technique of the BD has no gold standard[5]. The surgical approach has evolved from drainage procedures to the excision of the cyst and reconstruction of the bile duct as the treatment

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of choice[6]. There are two reconstruction techniques, hepaticojejunal anastomosis in Roux-en-Y and hepatico-duodenal anastomosis with no apparent benefit of any of them.

Materials and Methods

Twenty children with choledochal cyst managed in a pediatric surgical unit of CMH Dhaka with special interest in hepatobiliary and pancreatic surgery from March 2006 to February 2014 were reviewed. All the paediatric patients were <10 years of age. The presenting complaints, investigations, operative details and post- operative course were analyzed. Histopathological confirmation of the diagnosis was obtained in all cases. Follow-up information was obtained from outpatient records.

Results

There were 6(30%) males and 14 (70%) females with a male:female ratio of 1: 2.33. The incidence was highest between 7-10 years of age (Table-I).

Table-I: Distribution of patients according to age and sex (n=20)

Age (yrs)	Sex	
	Male	Female
>1-3yr	0	4
>3-5yr	2	3
>5-7yr	1	0
>7-10yr	3	7

In all, 85% of the patients presented with right upper abdominal pain, 70 % with history of jaundice and 20% of them presented with Palpable abdominal mass; vomiting in 70%, fever in 30% and hepatomegaly in 40% cases (Table-II).

Table-II: Distribution of patients according clinical presentation (n=20)

Clinical Features	No of Patient	%
Abdominal pain	15	85%
Jaundice	14	70%
Palpable abdominal mass	4	20%
Vomiting	14	70%
Fever	6	30%
Hepatomegaly	8	40%

*1 patient might have more than 1 features or symptoms

Serum bilirubin and alkaline phosphatase were elevated in 55% and 85%, respectively.

Table-III: Distribution of results of liver function test of patients (n=20)

Liver function test (LFT)	Number of patients	
	Normal	Raised
S. bilirubin	9(45%)	11(55%)
Alkaline phosphatase	3(15%)	17(85%)
SGPT	17(85%)	3(15%)

As a diagnostic approach, abdominal ultrasound was performed on all patients, supporting the CC diagnosis in 60% cases. Of the remaining eight patients, eight had a CECT scan, and two had MRI, to achieve the diagnosis of CC in all the eight patients.

Table-IV: Distribution of diagnostic approaches carried out on cases (n=20)

Test	No of patient	%
USG of HBS and pancreas	20	100
CECT scan of abdomen	8	40
MRCP	2	10

The different types of choledochal cysts as per Todani's classification were quite limited in the paediatric group. Among them 18(90%) had type I cysts and 2(10%) had type IV. In all 20 cases cholecystectomy, excision of choledochal cyst and Roux-en-Y hepaticojejunostomy were done, including those two patients who had type IV because of the broad base of the cyst.

Table-V: Distribution of types of choledochal cysts (n=20)

Type of Cyst	Number	Percentage
I	18	90
II	0	0
III	0	0
IV	2	10
V	0	0
Total	20	100

Post-operative complications were observed in 10(50%) cases. The early post-operative complications were observed in 4(20%) cases; fever in 2(10%) cases, pain and vomiting in one patient (5%), bile leakage in one (5%) case.

Table-VI: Distribution of cases with post operative complications (n=20)

Type	Complications	Number	%
Early	Fever	2	10
	Pain and vomiting	1	5
	Bile leakage	1	5
Late	Wound infection	2	10
	Recurrent cholangitis	3	15
	Anastomotic biliary leakage	1	5

Discussion

Choledochal cyst (CC) is a rare congenital anomaly of the bile duct[8], defined as a cystic dilation of the biliary tract[8-10], it's etiology is not well defined, Babbitt DP[11] theory is the most accepted etiology theory, proposing

that an abnormal junction between the biliopancreatic duct (ABPJ) create a shared channel that allows reflux of pancreatic secretion into the biliary tract thus causing increased pressure with subsequent ductal dilation, for which there is no standardized surgical treatment[9]. If left untreated CC has complications that range from BD obstruction to cholangiocarcinoma[12]. Further authors have found this association in up to 96% of cases[9,10,13]. Other congenital anomalies have been associated with CC like double common bile duct, sclerosing cholangitis, hepatic fibrosis, pancreatic cyst, annular pancreas and cardiac abnormalities[10]. In this study no congenital anomaly associated with CC was found.

Choledochal cysts are usually diagnosed in childhood, with 60% diagnosed before the age of 10 years, while only 25% are detected in adult life[14,15]. They also have an unexplained female: male preponderance[15], commonly reported as 4:1 to 3:1. International literature reports a 3–4:1 female to male ratio with predominance in Asian population[8-10]. In this study, there were 6(30%) males and 14(70%) females with a female:male ratio of 2.33:1.

The classical clinical triad of jaundice, pain, and a palpable mass can be found in about 20% of cases, with predominance in childhood[11,13]. In this study, this triad was found only in three patients which makes up for 15%. Jaundice and abdominal pain are the most dominant clinical features in children[13], these symptoms were found in our series in 70% and 85% respectively (Table-II).

In the study of Silva-Baez H et al paraclinical tests showed elevated liver enzymes (AST and ALT) in 92.8% of patients as well as elevated alkaline phosphatase in 100% and 57.1% had hyperbilirubinemia with an obstructive pattern[16]. But in this study serum bilirubin and alkaline phosphatase were elevated in 55% and 85%, respectively (Table-III). Liver function tests may be useful in narrowing the differential diagnosis. Hepatocellular enzyme and alkaline phosphatase levels may be elevated. None of these tests are specific for the diagnosis of a choledochal cyst.

A number of imaging modalities can be used to detect a choledochal cyst, such as ultrasonography, contrast enhanced computed tomography (CECT), MR cholangio-pancreatography (MRCP), endoscopic retrograde cholangio-pancreatography (ERCP) and percutaneous transhepatic cholangiography. MRCP, in particular, can be helpful in detecting an abnormal pancraticobiliary junction[17].

According to Miyano and Yamataka, the preferred initial radiologic examination in the diagnostic workup of a choledochal cyst is an abdominal ultrasonography scan[18]. Once a preliminary diagnosis is made using ultrasonography scanning, other supportive studies may be ordered, including abdominal CECT scans, MRI studies, or MRCP examinations[19]. These studies demonstrate the cyst with more precise anatomic detail.

In this study as a diagnostic approach, abdominal ultrasound was performed on all patients, supporting the CC diagnosis in 12 patients (60%) in other studies USG supported diagnosis of choledochal cyst in 50% and 63.4% cases respectively[20,21]. In this study, ultrasonogram report was suggestive for Choledochal cyst in eight cases (40%) and all of them underwent CECT scan in the present study. In other studies, CT-scan was performed in 5(50%) patients to sub categorize the type of choledochal cyst[20] and 9(21.95%) cases underwent CECT to subcategorize the type of CC as USG showed that gross dilatation of intrahepatic biliary tree[21]. Two(10%) had MRI for the confirmation of type of choledochal cyst in our study (Table-IV) and in another study they did MRCP on 3(7.31%) patients[21].

The most common are type I and IV and this preponderance was also seen in this study. Interestingly, the paediatric population only presented with type I and IVA, where as the adults present with almost full representations. The aetiology of type I and IVA cysts was thought to be due to an abnormal pancreaticobiliary duct junction, which results in a long common channel and mixing of pancreatic and biliary juice, leading to mucosal breakdown and dilatation[11].

In the study of Baez et al after confirmation of diagnosis of choledochal cyst by imaging studies, there were 9(64.28%) cases of type I and 5(35.71%) cases of type IV choledochal cyst thus treatment consisted of complete excision of cyst, cholecystectomy, and biliary tract reconstruction[22.] In this study, 18 patients (90%) had type I cysts and 2(10%) had type IV choledochal cyst (Table-V) and in all 20 cases complete excision of cyst, cholecystectomy, and biliary tract reconstruction were done. In commonest types of Choledochal cyst (Type I and IV), the treatment is surgery, although the reconstruction technique of the bile duct has no gold standard[5]. The surgical approach has evolved from drainage procedures to the excision of the cyst and reconstruction of the BD as the treatment of choice[6].

There are two reconstruction techniques, hepaticojejunal anastomosis in Roux-en-Y and hepaticoduodenal anastomosis with no apparent benefit of any of them.

Postoperative morbidity is 15-30%, mainly due to surgical wound infection, cholangitis and leakage from the anastomosis which in most cases are managed conservatively[23]. In this study, post-operative complications were observed in 10(50%) cases. In this study there was no postoperative haematoma or subhepatic collection, as seen in other study[23,24]. The early post-operative complications were observed in 4(20%) cases, fever in 2(10%) cases, pain and vomiting in one patient (5%), bile leakage in one (5%) case. All these patients were treated conservatively and recovered well.

Late complications were observed in 6(30%) cases. Wound infection in 2(10%) cases, recurrent cholangitis in 3(15%) cases and Anastomotic biliary leakage in one (5%) case. Wound infection in 10% cases was treated by regular dressing, in study of Jesudason SRB et al wound infection rate was 14.03% and settled by conservative treatment[25]. In this study anastomotic biliary leakage was 5%, reexploration was done and there was leakage from anastomotic site and it was repaired, in another study there were three anastomotic leaks (5.3%), two of which settled with conservative management. The third patient required surgical intervention[25]. There were no deaths during the first 30 days postoperatively, so there was no operative mortality, in study of Jesudason SRB et al. there were two postoperative deaths[25].

Conclusion

In infants with cholestatic jaundice and older children with intermittent abdominal pain in right hypochondrium should be evaluated for choledochal cysts. Ultrasonography of the abdomen is a basic investigation for evaluation of a patient of choledochal cyst. Most CC warrant resection in order to avoid future malignancies and future complications. Management includes total cyst excision and bilioenteric reconstruction.

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