ORIGINAL ARTICLE

Management of Choledochal Cysts- 7 years experience

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ABSTRACT

Aim: To study presentation and management of Choledochal cyst, its complications in children.
Methods: We performed a retrospective review of records of patients having choledochal cyst, (2006-2013) which were presented to Sheikh Zayed Hospital, Rahim Yar Khan, during last 7 years. We included clinical data, presentation, surgical management, post-operative complications (both early and late) and follow up data and analyzed and outcome recorded.
Results: There 14 patients in total out of which 6 were male and 8 were female patients. Mean age of presentation was 7 years, the range being from 2 months to 18 years. The incidence was highest in 5-6 years. In our series 71% of the patients presented with pain right hypochondrium, 29% presented with jaundice, 29% presented with a palpable mass in right upper quadrant of abdomen and 14% presented with hepatomegaly. In 4(28%) of the patients initially external drainage was done then definitive surgery was done after optimization of the patients. In all patients Choledochal cyst was excised and hepaticojejunostomy was done. Our survival rate was 100% and complication rate- 21%. In 2(14%) Choledochal cyst was adherent to portal vein and was excised by Lilly's technique. In 1(7%) there hepatic ducts more than 2, in that patient anastomosis leak was detected on 4th post operative day and was referred to a higher centre where re-anastomosed and survived. In 1(7%) minor wound infection was recorded at 5th post operative day.
Conclusion: Choledochal cyst is an important cause of jaundice with mass in right hypochondrium in neonates and recurrent right hypochondriac pain in older children. This disease should be kept in mind with high index of suspicions during examination of pediatric patients with above mentioned symptoms. Early referral of these patients may guarantee a better outcome.
Keywords: Choledochal cyst, hypochondrium, hepatomegaly

INTRODUCTION

Choledocal cysts is a congenital anomaly of bile duct which may involve intrahepatic biliary channels, extrahepatic biliary channels or both. Choledochal cyst is a rare congenital clinical condition of pediatric age group which requires early surgical intervention. It usually presents in infancy but sometimes its presentation is delayed till adulthood[1,3]. This is a benign condition[4], but may present with biliary tract malignancy in adulthood[17,23'25,27,28].

In literature these are also known as bile duct cysts, biliary cysts, congenital cystic dilatations of bile duct, but Choledocal cyst is more familiar to authors. Choledocal cysts are generally classified into 5 types[1,2]. This disease usually presents with jaundice, abdominal mass and abdominal pain[5]. Surgical treatment of Choledochal cyst is demanding and depends upon its type[6].

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MATERIALS AND METHODS

We are presenting 7 years (2006-2013) data of 14 patients who were presented to pediatric surgical ward in Sheikh Zayed Medical College/ Hospital, Rahim Yar Khan. We collected all data retrospectively. We reviewed all data regarding their clinical presentation, investigation findings, operative procedures, post-operative course, histopathological reports and follow up. All patients underwent a detailed clinical examination, ultrasonography of hepatobiliary tract, CT scan of biliary tract and MRCP when indicated. A complete baseline and follow up LFTs (including direct and indirect bilirubin) and coagulation profiles were obtained. All patients were optimized for surgery.

RESULTS

There were total 14 patients presented to us, out of which 6 patients were male and 8 were female patients, with male to female ratio of 1:1.33. The mean age of presentation was 7 years, the range being from 2 months to 18 years. The incidence was highest in 5-6 years of age.

Table of age distribution

Parameter	Results
Male to female ratio	1:1.33
Mean age of presentation	7 years
Range of age presentation	2 months to 18 years
Age of highest incidence	6-7 years

The cysts were classified according to Todani classification. All the patients were having fusiform dilatation i.e., type I. Ten patients having symptoms off and on pain right hypochondrium and four patients presented with jaundice and history of recurrent cholangitis. In four patients a mass was palpable in right hypochondrium and epigastrium. Two patients were having hepatomegaly.

Presentation of choledochal cyst

Clinical presentation	%age
Pain Rt. Hypochondrium	71
Jaundice	29
Palpable mass in RHC	29
Hepatomegaly	14

Liver function tests (LFTs) were normal in 20% of the patients, while serum bilirubin and serum alkaline phosphatase were elevated in 30% and 50% patients respectively. All patients underwent ultrasonographic scanning of hepatobiliary tree as the preliminary investigation. ERCP and HIDA scan were not performed due to unavailability. CT scan of hepatobiliary system was performed in all patients. In 01 patient multiple calculi were found in Choledochal cyst and gall bladder. In our series, 04 patients were inoperable due to severely deranged liver function tests and coagulation profile which were managed initially by external drainage procedure and definitive procedure was carried out after optimization of patients. 2 patients were having densely adherent Choledochal cysts with portal vein and excised by Lilly's technique.

Intraoperative findings

Intraoperative Finding	%age
Type of Choledochal cyst (type I)	100
Chol. Cyst adherent to portal vein	14
Presence of caliculi in chol. Cyst	7

Two patients were having more than two hepatic ducts at the portahepatis.

Post-op complications

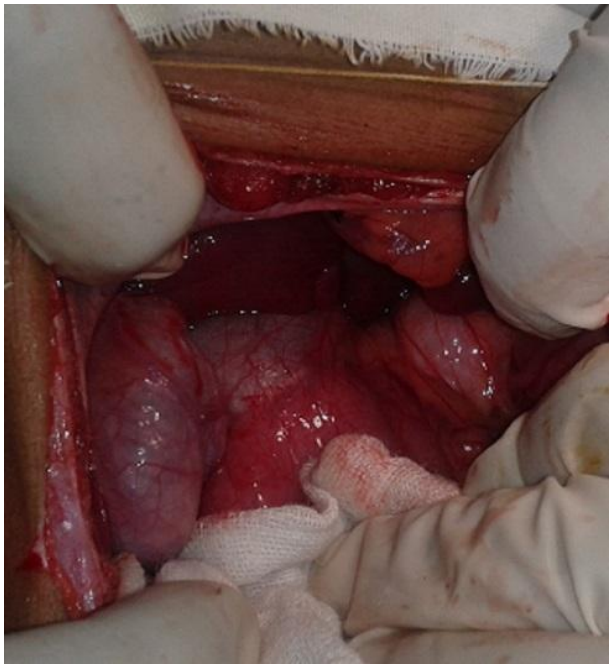
Complication	%age
Cholangitis	7
Minor wound infection	7
Anastomosis leak	7

In 01 patient occurred wound infection of mild severity which improved spontaneously with antibiotics.

One patients having multiple hepatic ducts developed anastomotic leak that was referred to a higher center where re-anastomosed and recovered. There was no post-operative mortality in our series. All the patients except one had complete recovery. Ten patients were available for follow up and remained symptom free after a mean follow up of 12 months. One patient having multiple hepatic ducts was having complaint of pain abdomen and was diagnosed as a case of cholangitis and managed conservatively.

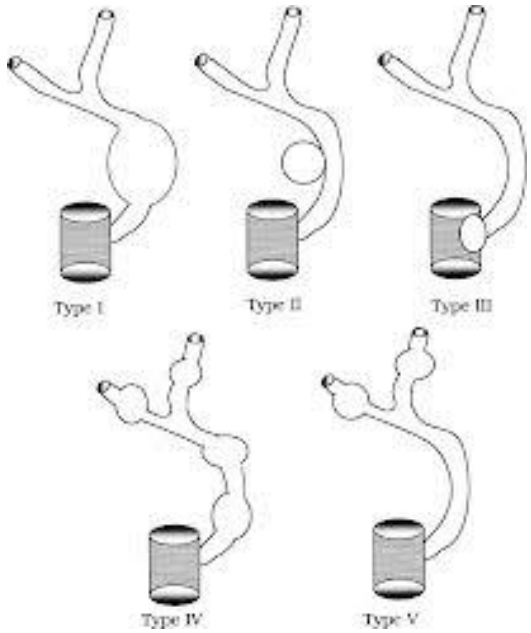
Post-operative results

Result	%age
Survival rate	100
Complete recovery	93



Todani classification of choledochal cyst

Type of Choledochal cyst	Description
Type I	Fusiform dilatation of extrahepatic bile ducts
Type II	Extrahepatic bile duct diverticulum
Type III	Choledochocele
Type IV	Dilated intra- and extrahepatic bile ducts
Type V (Caroli's disease)	Cystic dilations of intrahepatic bile ducts.



DISCUSSION

Choledochal cyst was first described by Vater[7]. Choledochal cyst is mainly a disease of childhood, but it may present adulthood in about 20% of the cases[6]. It was classified into 3 types initially Alonso-Lej et[8] and then by Todani into 5 types[9], however revised classification system has also been proposed[10] yet not widely accepted. Distribution of different types of cyst is as follows: 50%–80% are type I, 2% type II, 1.4%–4.5% type III, 15%–35%, type IV and 20% type V[12].

The incidence of Choledochal cyst is about 1:150000[11] with female predominance (~80%). The aetiology of the disease is still unknown.

Most widely used hypothesis used hypothesis is presence of an anomalous pancreatico-biliary ductal confluence (APBDJ) which predisposes the pancreatic enzyme reflux and subsequent deconjugation of bile acid. Activated pancreatic enzymes and deconjugated bile salts provoke a chronic inflammatory reaction at that site which weakens portions of biliary tree. Further pooling of deconjugated bile salts and activated pancreatic enzymes in weakened Choledochal cyst wall causes abnormal dilatation of Choledochal cyst. This abnormally dilated Choledochal cyst sometimes presents as mass in right hypochondrium and epigastrium[6,12,27]. Oligoganglionosis in the terminal portion of CBD causing proximal dilatation and functional obstruction is another hypothesis[13,27]. however still no hypothesis has been accepted.

Pattern of presentation of Choledochal cyst varies in patients. It may present with symptoms like upper abdominal pain, features of obstructive jaundice or upper abdominal mass. Classical triade of abdominal pain, jaundice and upper abdominal mass only in one third of the patients[14]. Presentation with two or three classic signs and symptoms is more likely in children (82%) than in adults (25%)[15]. Presentation in adults varies as compared to children as the clinical presentation is vague in adults and they are more likely to present with complications [16]. Infants are more likely to present with jaundice and clay colored stools. They often have palpable upper abdominal mass and hepatomegaly. Children after infancy are more likely to present with intermittent jaundice and upper abdominal pain, but mass still may be palpable. Adults are more vulnerable to develop complications and present with cholangitis or pancreatitis.

Antenatal diagmosis can be made on in 26th week[30,31] which may help in making arrangments of early surgical procedure.

Ultrasonography provides adequate information regarding intra- and extrahepatic biliary channels and liver size and texture. It is non-invasive, cheap and widely available but is operator dependent and a good sinologist is necessary for diagnosis in difficult and equivocal cases.

ERCP defines more accurate anatomy of biliary tree, but is invasive and associated with complications and expertise in children is also widely available. Added benefit of ERCP is that it can be therapeutic in some clinical situations.

MRCP being a non- invasive procedure is emerging technique and is an alternative to ERCP, but has no therapeutic option as compared to ERCP.

Demonstration of stagnation of bile by HIDA scan in the absence of biliary obstruction has been reported as diagnostic sign of Choledochal cyst.

Children may present with advance disease, usually malnutrition and coagulopathy aggravates the situation and makes definitive procedure difficult to be carried. Adults are more likely to present with complications like cholangitis, obstructive jaundice, pancreatitis, cholelithiasis, hepatolithiasis, intrahepatic abscesses, portal hypertension, biliary cirrhosis and cholangiocarcinoma.

Treatment option is more dependent on the type of Choledochal cyst. Complicated cyst management involves initial external drainage and then definitive procedure after optimization. Ideal treatment for type I and IV is complete excision and restoration of biliary-enteric communication by Roux-en-Y hepaticojejunostomy. In certain types if the disease is limited to resectable portion of liver then surgical treatment options like segmentectomy, sectionectomy and hemihepatectomy may be considered. If the cyst is adherent to portal vein then

it may be treated with Lilly's technique. In type V liver transplant may be a better option.

In recent years in pediatric patients, laparoscopic resection of congenital choledochal cyst and choledochojejunostomy is reported to be safe and effective and getting popularity with promising long term results, but considerable longer learning curve is a hindrance in clinical practice[18,19,32].

Miyano T et al in 1996 reported mean age of patients with Choledochal cyst at the time of surgery 4.3 years and development of intrahepatic bile duct stones with or without cholangitis in 2.3% of the cases after a mean follow up of 11 years[24], but in our series mean age of presentation was 7 years and incidence of cholangitis was observed in 01 patient (7%) after a mean follow up of 12 months. No patients developed intrahepatic bile duct stones in our series, however a long follow up is necessary to report development of intrahepatic bile duct stones.

Yamatka A et al. in 1997 reported in a series of 200 patients of pediatric age group that the mean age at which patients having Choledochal cyst became symptomatic was 3 years, mean age at the time of surgery was 4.2 years. In 88% of the patients a definitive procedure was done primarily and in 12% of the patients definitive procedure was done secondarily. They reported a complication rate of 9% and reoperations were done in 7.5% of the patients[26]. In our series mean age of presentation was 7 years and definitive surgery was done after a mean of 01 month of presentation. We did definitive procedure in 71% of the patients primarily, and in 29% of the patients definitive procedure was carried out after initial external biliary drainage. Our complication rate was 21%. In 7% of the patients a reoperation was done which correlates with the international statistics.Discordance in our statistics may be due to small number of patients in out study. Cvetkovic A et al. reported in 2011 reported a male to female ratio of 1:4[27], while in our study it was 1:1.33.

AL-Raymoony A, Samawi B in 2006 reported a male to female ratio about 1:1 (44% males: 56% females) in a series of 9 patients[29].

Lee KH et al. in 2009 reported a presentation of Choledochal cyst with abdominal pain (69%), jaundice (40%) and abdominal distension (4%). According to them complication rate was 19% in their study[32]. In our study abdominal pain was present in 71%, jaundice in 29% and abdominal distension in 29% of the cases. Our complication rate was 21%.

CONCLUSION

Choledochal cyst is a rare but important clinical entity and should be considered in all patients with biliary colic, recurrent cholangitis or pancreatitis.

Approach to the patients with above mentioned symptoms should be systemic and a high index of suspicion should be in mind in order to diagnose Choledochal cyst. Management of the patients with Choledochal cyst is demanding and depends upon its type. Recently, laparoscopic resection of congenital choledochal cyst and choledochojejunostomy is being practiced in management of Choledochal cyst in pediatric age group.

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