ORIGINAL RESEARCH PAPER

Volume-8 | Issue-6 | June-2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CHOLEDOCHAL CYST: CLINICAL PRESENTATION AND MANAGEMENT, TWELVE YEARS' EXPERIENCE IN A TERTIARY CARE CENTRE.

General Surgery

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ABSTRACT

Background: Choledochal cyst is a rare congenital anomaly and the surgical intervention is mandatory which depends upon the type of the cyst.
Aim: To study the clinical presentation, management and post-operative complications after different operative procedures for choledochal cyst.
Methods: Forty patients were included in this retrospective study, out of which 26 were females and 14 were males. All patients were fully evaluated by clinical examination and all necessary investigations were done. 36 patients had Type –I CDC and 04 patients had Type –IV CDC. 21 patients underwent hepaticoduodenostomy (HD), 16 patients underwent hepaticojejunostomy (HJ) and 03 patients underwent hepaticoduodenostomy with appendicular interposition (HAD).
Results: We have survival rate of 97.5 % and complete recovery of 94 % with a single mortality in our study.
Conclusion: CDC is a rare congenital anomaly and its management is very important. Hepaticoduodenostomy is more physiological than hepaticojejunostomy and nowadays laparoscopic management is preferred.

KEYWORDS

Choledochal cyst (CDC), Hepaticoduodenostomy (HD), Hepaticojejunostomy (HJ), Hepaticoduodenostomy with appendicular interposition (HAD).

Introduction

Choledochal cyst (CDC) is a rare congenital anomaly of either extra hepatic or intrahepatic biliary tree or both.[1] It is a rare pathological condition of paediatric age group and early surgical intervention is mandatory. CDC usually presents in early infancy but it may have delayed presentation in adulthood.[2,3] There are 5 types of CDC.[2] CDC usually presents with jaundice ,abdominal mass and pain.[4] Surgical management of CDC is demanding and depends upon the individual type of cyst.[5] Its incidence is higher in Asian population 1 in 1000 with a male to female (M:F) ratio of 1:3-4.[6] Complete excision of the cyst is very important to avoid long term complications like recurrent cholangitis, gall stones and malignant transformation[7].

Methods

We are presenting a data of 40 patients who presented to our tertiary care hospital between 2007-2018 over a period of 12 years. We collected and reviewed the data retrospectively regarding their clinical presentation, clinical findings, investigations, operative procedures, postoperative course, histopathological reports and follow up. All patients underwent a thorough and detailed clinical examination. All routine investigations were done including complete hemogram, serum electrolytes, KFT, baseline LFT and Coagulation profile. Diagnostic investigations including Ultrasonography of hepatobiliary system, CT scan of biliary tract and MRCP when indicated. All patients were optimized properly before surgery. Regular follow up was done and patients were assessed by clinical examination, LFTs including direct and indirect bilirubin and coagulation profile and ultrasound as and when needed.

Results

Forty patients were included in this study, out of which 26 were females and 14 were males as shown in table -1. The age ranges between 3 months to 18 years and the mean age of presentation was 7.3 years. Commonest age group was 7-8 years.

Table 1. Age distribution of patients

Parameter	Result
Male to Female ratio	26: 14
Age (range)	3 months to 18 years
Mean age	7.3 years
Age of highest incidence	7- 8 years

Majority of the patients present with pain Right hypochondrium followed by jaundice, abdominal mass and hepatomegaly as shown in table 2.

Table 2. Clinical presentation of CDC.

Presentation	Percentage(% age)
Pain right hypochondrium	78
Jaundice	38
Abdominal mass	40
Hepatomegaly	18

Liver function test were normal in 22% of the patients and serum bilirubin was raised in 60% of the patients and 56% had raised serum alkaline phosphatase. All patients undergo USG hepatobiliary system as first line investigation. CT scan was performed in all patients and MRCP in 34 patients. HIDA scan and ERCP were not available and hence not performed in any patient. In 02 patients cholelithiasis and stones in the cyst was seen. In 03 patients the CDC was densely adherent to the portal vein and was excised by Lilly's technique. Intraoperatively it was found that 36/40 had type -1 CDC and 4/40 had type- IV CDC and none of the patient had type II, III and V CDC as shown in table- 3. The cysts were classified as per Todani classification table 4.

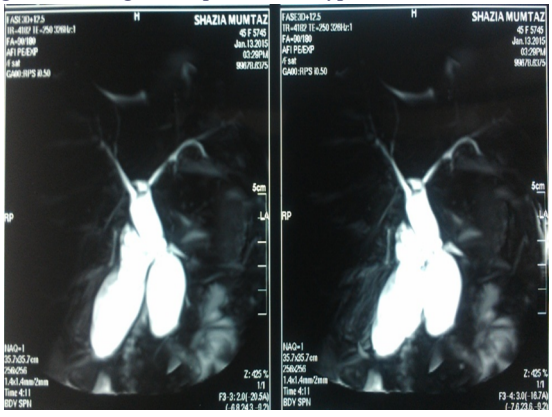
Table 3. Intraoperative findings

Type of CDC	No. of patients
Type I CDC	36
Type II, III	Nil
Type IV	4
Type V	Nil

Table 4. Todani classification of choledochal cyst.

Type of CDC	Description
Type I	Fusiform dilatation of extrahepatic ducts.
Type II	Diverticular dilatation of extrahepatic ducts.
Type III	Choledochocele.
Type IV	Both intra and extrahepatic bile duct dilatation
Type V (Carolis disease)	Intrahepatic duct dilatation.

Figure 1 showing MRCP picture with Type –I CDC.



Out of forty patients 21/40 underwent hepaticoduodenostomy (HD), 16/40 underwent hepaticojejunostomy (HJ) and three patients underwent hepaticoduodenostomy with interposition of appendix (HAD) as enteric interposition as shown in table 4. Since HAD is mentioned in literature, we did it in 03 patients initially.

Table 5. Operative procedures done

Operative procedure	No of patients
Hepaticoduodenostomy (HD)	21
Hepaticojejunostomy (HJ)	16
Hepaticoduodenostomy with appendix interposition (HAD).	03

Figure 2. Intraoperative picture showing gall bladder, CDC and common hepatic duct.

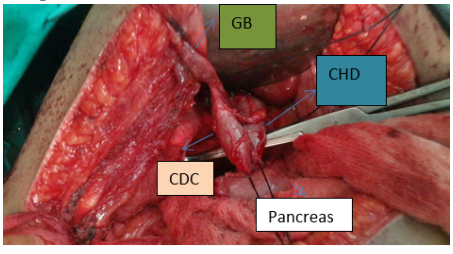


Table 6. Postoperative complications.

Early 6	Late 7
Bile leak -4	Cholangitis -2
Pancreatic fistula -1	IHD stones – 2
Death -1	SAIO -3

Postoperative complications in our series are shown in table 5. We have one mortality in our series. 24 patients who came for follow up were followed over a mean period of one year and remained symptom free. One patient with multiple hepatic ducts had multiple episodes of cholangitis and was managed conservatively. The overall survival rate was 97.5% and complete recovery was seen in 94% of patients.

Discussion

It was Vater who first described the CDC[8]. CDC is usually a disease of infancy and childhood but 20% may present in adulthood.[5] Todani the accepted classification , has classified the CDC into 5 types.[9] About 50 - 80% are type I, 2% are type II, 1.4-4.5% are type III, 15-35% type IV, and 20% are type V.[10] The most widely accepted theory regarding the aetiology of CDC is anomalous pancreatico –biliary ductal junction confluence (APBDJ) which predisposes pancreatic enzyme reflux and de conjugation of bile acids which leads to chronic inflammatory reaction and weakening of biliary tree wall as shown in figure-3.

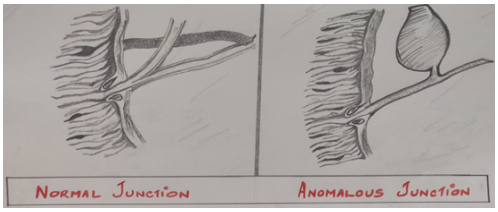


Figure 3. Showing abnormal pancreatico-biliary duct junction confluence

Classical triad of abdominal pain, jaundice and upper abdominal mass is present in one third of patients.[11] This classical presentation is more common in children 82% than in adults 25%.[12] Antenatal diagnosis can be made at 26 weeks of gestation and it allows to make arrangement for early intervention.[13, 14] Ultrasound gives adequate information regarding the nature of the cyst like whether it is in biliary or extra biliary, size of the cyst and texture. ERCP is more accurate and therapeutic but since it is invasive and is associated with complications, MRCP is preferred. HIDA Scan can be done which can show stagnation of bile in absence of biliary obstruction is a diagnostic sign of CDC. Some children may present in later stage of life with malnutrition and coagulopathy. Adults may present as a case of obstructive jaundice, pancreatitis, hepatolithiasis, hepatic abscess, portal hypertension, cirrhosis and cholangiocarcinoma. Treatment of complicated cyst needs external drainage initially followed by definitive procedure. Definitive treatment depends upon the individual type of cyst. For type I and IV, the ideal treatment is excision of cyst with biliary –enteric anastomosis by Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy. In type V liver transplant may be needed. In type II endoscopic management can be done. If the cyst is adherent to portal vein Lilly's technique can be done. In recent years laparoscopy has evolved in the management of CDC. [15,16] In a study by Miano et al[17] the mean age was 4.3 years and the development of intrahepatic stones with or without development of cholangitis was 2.3% after a mean follow up of 11 years. In our series the mean age of presentation was 7.3 years and cholangitis was observed in 02 patients over a follow up period of 12 months. No patient developed intrahepatic stones; however a long follow up is needed. Lee KH [18] in their series had a presentation of abdominal pain 69%, jaundice 40% and abdominal mass in 4% and the complication rate was 19%. In our study the presentation was like pain in 78%. Jaundice 38% and abdominal mass 40%. Our complication rate was 32.5%.

Conclusion.

Choledochal cyst CDC is a very rare anomaly and should be suspected in any patient with recurrent attacks of cholangitis, pancreatitis and obstructive jaundice. As for as management is concerned, hepaticoduodenostomy is more physiological than hepaticojejunostomy and the latest trend is laparoscopic management by laparoscopic choledochojejunostomy.

Conflict of interest. None

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