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Original Research Article

Congenital choledochal malformations in children: management strategies

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ABSTRACT

Background: A Congenital Choledochal Malformations (CCM) is common congenital defect of biliary tree in Asian subcontinent. Presentations of CCM are vague from asymptomatic to life threatening cholangitis or pancreatitis. Complete cyst excision and bilioenteric anastomosis is now accepted surgical treatment.
Methods: This is a retrospective study of five years duration. In this study the clinical presentation, haematological, biochemical and radiological findings, operative procedure and outcome were studied from case records.
Results: Total of 20 patients were studied, with clinical presentation of recurrent abdominal pain in eight cases, previous history of cholangitis in five cases, acute cholangitis two cases, biliary peritonitis in four cases and previous history of pancreatitis in one case. In 13 cases Roux-en-Y hepaticojejunostomy (RYHJ) and in 4 cases hepaticoduodenostomy (HD) done after cyst excision. One case of type II CCM managed with only cyst excision, in two cases Lilly's procedure, one case requires temporary external drainage and in two cases temporary internal drainage done in view on cholangitis and jaundice.
Conclusions: However, the definitive treatment of CCM is complete cyst excision and bilioenteric anastomosis, though lot of other temporary majors are also required at different stages of disease.

Keywords: Congenital Choledochal Malformations (CCM), Hepaticoduodenostomy (HD), Roux en Y Hepaticojejunostomy (RYHJ)

INTRODUCTION

The incidence of Congenital Choledochal Malformations (CCM) in Asia is 1/1000 live birth.[1] The exact incidence in India is not known. CCM is more common in females with a ratio of 3:1. Presentation in children is very nonspecific and vague. The classical presentation is triad of symptoms pain abdomen, palpable mass in right upper abdomen and jaundice. Although abdominal ultrasound scan (USG) is the first and easily available diagnostic modality but Magnetic resonance cholangiopancreatography (MRCP) is the "gold standard" for the diagnosis and planning the management. Contrast enhanced computed tomography (CECT) abdomen is also helpful in cases present with abdominal mass. Endoscopic retrograde cholangiopancreatography (ERCP) is an invasive investigation and require expertise in children.

The most commonly followed classification for CCM was initially given by Alonso-Lej et al. in 1959 and later Todani et al. in 1977 modified it.[2,3] Lilly et al. described and Sarin et al. give clinical significance of an entity called "forme frusta.[4,5] The most commonly proposed theory for CCM is supposed to be caused by an anomalous pancreatic-biliary junction. Complications of CCM include pancreatitis, cholangitis, secondary biliary

cirrhosis, spontaneous rupture of cyst, and rarely cholangiocarcinoma.

Treatment of CCM is complete excision of the cyst and bilioenteric anastomosis. A lot of other interventions are also needed to manage the disease at different stages like external drainage (need the help of intervention radiologist), sometime temporary endoscopic stenting (need the help of gastroenterologist expert in paediatric cases), peritoneal lavage and T tube or a sub hepatic drainage in cases of biliary peritonitis.

METHODS

This is a retrospective study done of last five-year durations from January 2009-December 2014. Patient's demography, clinical, biochemical findings, radiological findings, surgical procedure and outcome were recorded. The diagnosis of CCM was established by either USG, CECT abdomen or/and MRCP. In those presenting with acute symptoms, they were tided over the acute episodes before definitive surgical intervention with intravenous third generation cephalosporin and aminoglycoside. In those cases, whose acute inflammation did not settle on conservative treatment, an ERCP with/without a papillotomy and CBD stenting would be performed.

In cases where ERCP stenting failed or not possible and still the child was not responding with antibiotics, percutaneous transhepatic drainage of biliary system were done. For type I CCM, our standard treatment is complete cyst excision and with a RYHJ or HD. We prefer to do HD in cases where diameter of hepatic duct was less than 6 mm after cyst excision and rest of cases we are routinely doing RYHJ. The similar principle was applied to type IVa cysts initially but a more radical excision of the bile duct to the hilum was performed to remove any narrow area at confluence of CBD. For cases of type II CCM only cyst excision was practiced. The Lilly's mucosectomy would be used if the cysts were densely adhered to the adjacent vascular structures making complete excision dangerous.

In cases of perforated CCM with biliary peritoneal collection, our initial approach was to drain the collection and give time to settle the inflammations. In this study the management strategies, either temporary for short term symptomatic improvement (to improve the general condition so that patient can undergo major surgical procedures) or definitive surgical procedures were studied. Early postoperative complications like 24 hours fever monitoring and total and deferential leucocyte count were recorded on daily basis.

In this study we also tried to find out the relation of intraoperative hepatic duct saline flushing and features of cholangitis (fever spikes above 100 ºF and leukocytosis). Long term follows up of patients recorded from last follow up visit or telephonically whichever were possible.

RESULTS

Total of 20 cases of CCM were managed by us in five-year durations. Male to female ratio were 9:11, age ranges were 1 year to 12 years (median age was 5.5 years). Presentation were recurrent abdominal pain in 8 cases (including four cases of recurrent abdominal pain with palpable abdominal mass), previous history of cholangitis in 5 cases, acute cholangitis in two cases, biliary peritonitis in 4 cases and previous history of pancreatitis in one case (Table 1). In one patient who presented with biliary peritonitis had left lobe small liver abases (6-8 milliliters).

In all patient initial evaluation were done with USG abdomen. MRCP done in 8 patients only (as these require anaesthesia) and CECT abdomen done in 7 cases. ERCP stenting done in two cases presented as acute cholangitis, as these were not responding to antibiotics. One case requires temporary external drainage with trans-hepatic pig tail catheter drainage of biliary system, as endoscopic stenting failed in this case and child was not fit for anaesthesia for cyst excision at that time. In 13 cases type- I, one case type II, in two cases type IVa, two cases with type IVb, in one case type V and in one case forme fuste of CCM were found (Table 1). We are doing open surgery for CCM. Our technique of CCM surgery was exploration of right upper abdomen by subcostal incision. After initial evaluation of the anatomy of common bile duct (CBD), liver mobilized.

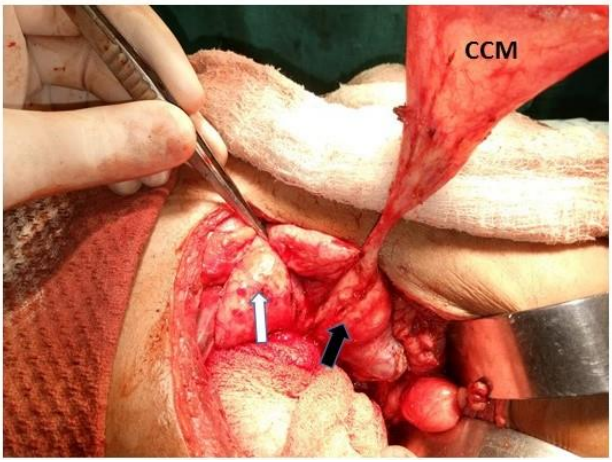

Figure 1: Dissected intra-pancreatic lower part of CCM (white arrow showing duodenum and black arrow showing pancreas).

After small dissection at Calot's triangle cystic artery was ligated. After mobilization of gall bladder from liver bed, it was used as traction to mobilize the CCM. Cyst separated carefully posteriorly from portal vein and hepatic artery taking care of variation of the anatomy. On an infant feeding tube cyst is transacted, upper end mobilized and transacted just 5 mm below confluence of hepatic duct or depending on the anatomy. In cases where intrahepatic dilatation of biliary ducts occurred (like Type

IVb CCM) our principle was correction of distal stenosis at the confluence of hepatic duct and bilioenteric anastomosis. Lower part of cyst removed, intrapancreatic after mobilisation of the duodenum, and divided just 2-3 mm above the opening of pancreatic duct taking the imaging study as a roadmap (Figure 1).

Table 1: Clinical features, types of CCM (radiological findings), approach of management and outcome.

Presentation	Number of cases	Type of CCM	Managements strategies	Any significant event/complications
Recurrent pain abdomen and/or palpable mass	8	1 forme fuste, 6 type I and 1 type IVa	RYHJ done in six cases, including one done after Lilly's procedure. HD done in two cases including one done after Lilly's procedure.	One patient develops minor bile leaks of type I CCM in which after cyst excision RYHJ was done
History of cholangitis	5	2 type I, 1 type II, 1 type IVa and 1 type IVb	Three cases were managed with cyst excision and RYHJ including one case that temporary managed with external drainage, one case managed with cyst excision and HD and one case of type II CCM managed only with cyst excision	All patients were doing well in follow up
Acute cholangitis	2	1 type V and 1 type IVb	Managed with antibiotics and endoscopic ERCP stenting of CBD	One case with type V CCM were lost from follow up, other were managed with cyst excision and RYHJ
Biliary peritonitis	4	All type I	In two cases lavage of peritoneum and T tube drainage of CBD, in one case lavage of peritoneum and sub hepatic drainage and in one cases cyst excision and RYHJ as a primary surgery done	Two cases that managed with T tube drainage of CBD were latter cyst excision and RYHJ done and a case that primary surgery of cyst excision and RYHJ done were doing well in last follow up, cases that managed with lavage and sub hepatic drain was lost from follow up.
History of pancreatitis	1	I	Cyst excision and HD	Minor bile leaks in drain

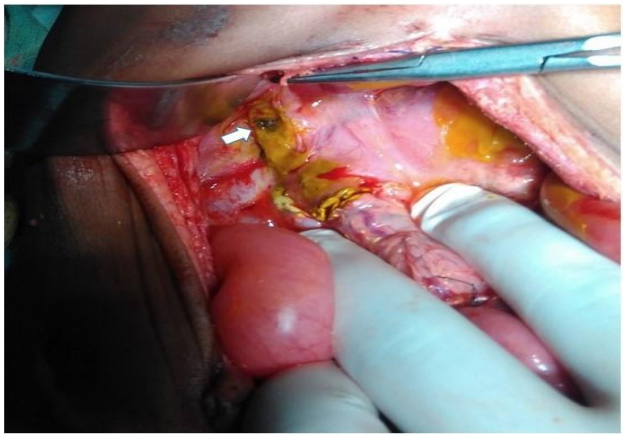

Figure 2: Intraoperative view of perforated CCM, perforated site on anterior wall of cyst (shown by arrow).

CBD at biliopancreatic junction were flushed with copious saline to clear any debris, mucus or stone. Proximal intrahepatic duct flushing was done on cases to case basis. In 13 cases RYHJ and in 4 cases HD done after cyst excision, one case of type II CCM managed with only cyst excision, in 2 cases complete cyst excision were not possible due to sever adhesion so Lilly's procedure was done.[6] Four cases who presented with biliary peritonitis (Figure 2), T tube placed in CBD after lavage in two cases, sub hepatic drain placed in one case and excision of cyst and RYHJ done in one case. One case of biliary peritonitis in which sub hepatic drain was placed, definitive procedure was not done, as he lost from follow-up. In two cases (one of type IVb and another type V CCM) endoscopic stenting of CBD done in view of cholangitis not responding to usual conservative treatment, however definitive treatment of cyst excision was done in only one (one case of type V CCM lost from follow up). Two patients developed minor biliary leaks in sub hepatic drain, improved on conservative management. In total of nine cases intraoperative intrahepatic bile duct saline flushing were done. Six cases of them were developed fever in postoperative period and

four they had leukocytosis (Table 2). These cases who had fever and leukocytosis required up gradation of antibiotics. Post-operative stay at hospital were 5 days to 37 days (average 11.6 days). In follow, up of 8 months to 2½ years no complications occurred in our cases.

Table 2: Features of cholangitis (fever and leukocytosis) after intraoperative saline flushing of hepatic duct.

Intra operative flushing of hepatic duct	Cholangitis in post-operative period		Total cases
	Yes	No	
Yes	6	3	9
No	1	8	9
Total cases	7	11	18

DISCUSSION

The exact aetiology of CCM remains unknown but the "common channel theory" as proposed by Babbit in 1969 has been popular.[7] As per this theory abnormal pancreatic biliary junction resultant pancreatic juice reflux into the biliary tree causes ductal changes. Abnormal function of the sphincter of Oddi has been also reported to predispose to pancreatic reflux into the biliary tree and an association has been seen with increased sphincter of Oddi pressures and spasm.[8] Inadequate autonomic innervations demonstrated by Kusunoki et al that suboptimal number of ganglion cells in the narrow portion in the common bile duct in patients with a CCM, as compared with controls suggesting a functional obstruction.[9] CCM have a wide range of presentation contributing to the diagnostic challenge in some of these patients.[10] Vague clinical presentation of recurrent abdominal pain was found in majority of our cases. This presentation may pose diagnostic difficulty if not associated with palpable abdominal mass. Patients frequently have only intermittent attacks of colicky abdominal pain if they have elevated amylase and lipase concentrations lead to the proper diagnostic workup. However, some of patients presented acutely with abdominal pain with or without cholangitis, obstructive jaundice. Though the other presentations of CCM are pancreatitis, and rarely biliary peritonitis and liver abscess.

In our study four cases presented with biliary peritonitis and one cases of biliary peritonitis also had small left lobe liver abscess. The CCM perforation was first described by Weber in 1934, only 56 cases were reported till date and incidence of this was only less than 2%.[11-13] No definite cause could be ascertained in a majority of cases of CCM perforation. Anomalous biliopancreatic duct junction has been implicated in few cases. It is commonly accepted that it occurs due to reflux of pancreatic juices resulting from an abnormal biliopancreatic ductal junction along with mural weakness. Perforation of the common bile duct may occur due to the abrupt increase in intraluminal pressure due to obstruction by protein plugs at the common channel that results in decreased blood flow in the vessels which run along the lateral border of the bile ducts resulting in ischemia on the anterior surface of the bile duct.[14] The onset of symptoms in our cases were acute in one cases and subacute in rest of three cases. In acute forms child presented with vomiting, severe abdominal pain and shock. Rest of three cases presented in subacute form as 'biliary ascites' and had acholic stools, mild fluctuating jaundice and ascites. Definite diagnosis of the entity is difficult, due to these cases usually lacking a history suggesting a CCM, collapse of the cyst following perforation may prevent its identification on radiological studies. In our cases diagnosis was made on strong clinical suspicions, elevated serum bilirubin, presence of bilirubin on ascitic fluid analysis and in one cases technetium 99 sequential scintiphotography helped us in the diagnosis.[15] Initial treatment of biliary peritonitis consists of drainage of the extrahepatic biliary system by cholecystostomy or T-tube choledochotomy. The definitive procedure is best delayed till later. In our case we drain the CCM with T tube in two cases, one cases managed with sub hepatic drain and one case, cyst excision and RYHJ was done as an initial surgery. We recommend that when a child comes with an acute presentation, T tube drainage at the perforation site should be done. However, when the child presents late with features of biliary peritonitis with friable and oedematous tissues, a simple drainage via a cholecystostomy is an excellent option, in an emergency setting.

Percutaneous biliary drainage performed in one of our case. Patient presented with jaundice, right upper quadrant pain, palpable mass and previous history of cholangitis. Later the child was managed with cyst excision and RYHJ. Percutaneous transhepatic biliary interventions can be performed safely and effectively as a bridge prior to surgery in benign diseases of children.

Two of our patients need endoscopic biliary stenting as they are presenting in stage of acute cholangitis and not responding to antibiotics, one of them was type V CCM and other type IVb. Both the cases were improved after the procedures, one of them of type IVb CCM was operated with cyst excision and RYHJ. Case of type V CCM that was initially managed with endoscopic biliary stenting was lost from follow up after improvement of initial acute cholangitis. ERCP is the most sensitive technique to define the anatomy of the biliary system, but can be difficult to perform in the pediatric population given the need for general anaesthesia. An ERCP allows for direct visualization of the pancreaticobiliary junction. In addition to its diagnostic yield, ERCP can be therapeutic by allowing biliary drainage and endoscopic sphincterotomy of choledochoceles (type III CCM). This procedure is associated with potential complications, including bleeding, cholangitis, acute pancreatitis, and perforation. As a result, this should be reserved in

paediatric cases when other methods are failed to relive the intra biliary pressure.

Excision of the CCM in presence of acute suppurative cholangitis has been reported to be associated with high morbidity and mortality. Staged management in the form of an initial biliary decompression followed by cyst excision 6-12 weeks after the control of cholangitis, has been found to give a satisfactory outcome.[16] All of our cases that were managed in staged intervention either percutaneous trans hepatic external biliary drainage, internal biliary stenting or cases of biliary peritonitis, definitive surgery of cyst excision and bilioenteric anastomosis were done at an average interval of 10 weeks.

One of our case had previous history of pancreatitis, managed with cyst excision and HD. The association of acute or chronic pancreatitis with CCM has been explained by an underlying anomalous biliopancreatic junction which permits free intermixing of pancreatic and biliary juices and protein plugs in the common channel.[17] The incidence of acute pancreatitis with CCM are variable, and true incidence is difficult to decipher because serum amylase or lipase levels are not routinely done for pain abdomen in patients with CCM.[18] In most cases, episodes of pancreatitis are mild and recurrences can only be eliminated by excision of cyst.[19]

The basic principle of definitive surgery is to excise the entire dilated extrahepatic bile duct without damaging the adjacent vital structures such as the portal vein, the hepatic artery, the pancreas and its duct and the duodenum. It is also important to clear up the protein plugs/sludge at the common channel before closure of the ductal opening. For the upper margin of transaction, it is usually chosen at the site of the common hepatic duct well proximal to the cyst and a wide anastomosis is advocated to avoid subsequent stricture. Lower end dissected intra pancreatic after small mobilisation of second part of duodenum taking the help of MRCP or CECT as a roadmap taking care to avoid injury to pancreatic duct. It is also important to ascertain the clearance of stones or sludge in the intrahepatic ducts and lower intra pancreatic part of ampulla, the remains of such will invite future troubles. In some cases, depending on surgeon preference or on radiological investigation if any hepatic duct debride or stone, saline flushing of hepatic duct after inserting a feeding tube inside done.

In literature the result of HD is variable. Some study like Shimotakahara et al. and Elhalaby et al. disfavours due to high duodenogastric bile reflux, while Silva-Baez et al and Mukhopadhyay et al, favours the procedure of HD due to shorter operative time and avoidance of intestinal anastomosis.[20-23] A recent meta-analysis study by Narayanan et al. support the procedure of cyst excision and RYHJ.[24] In our experience, we have done HD in five cases, in two of them developed post-operative minor bile leak in drain that improve on conservative measures no redo surgical intervention required. On follow up of these cases up to 2½ years, none of them developed complications of biliary gastritis, cholangitis or anastomotic site stricture. Our principle of HD was after cyst excision duodenum should be adequately mobilised, bilioduodenal anastomosis should be done at junction of first and second part of duodenum that is away from pylorus. In our cases there was no difference between RYHJ and HD on follow up.

In total of nine cases of CCM intraoperative intrahepatic bile duct saline flushing were done during definitive surgery after cyst excision. Six cases of them were developed fever in postoperative period and four of them had leucocytosis. These cases who had fever and leucocytosis, up gradation of antibiotics was done. There was high incidence of post-operative fever, leucocytosis and long post-operative stay in cases where intraoperative saline flushing of intrahepatic duct done as compared to cases in which intraoperative flushing not done. Our proposed theory to explain this phenomenon as there is some degree of dilation of intrahepatic biliary system was present in cases of CCM. In CCM due to bile stasis, there was some degree of chronic infection may present that will translocated proximally in liver during saline flushing that will leads to early postoperative complications of cholangitis. On this finding our recommendation is that saline flushing of hepatic duct should be done only in selected cases where previous suspicion or diagnosis of intrahepatic dust stone or slough was present otherwise in every case it should not be done. Though our sample size is small and this may have depended on several factors as individual anatomy, surgeons' technique and other factors.

CONCLUSION

Management strategy for complicated CCM needs meticulous planning and multimodality approach. The therapeutic strategy needs to be tailored to an individual case. Percutaneous and endoscopic biliary drainage procedures are useful adjuncts to the surgical management. The outcome is satisfactory in patients with appropriately managed complicated cases. Early surgical intervention should be done in view of sever life threatening complications of CCM. Intraoperative flushing of hepatic duct should be done in only selected cases due to high postoperative complication of cholangitis.

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