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Research Article

MANAGEMENT DILEMMA WITH CHOLEDOCHAL CYST IN PAEDIATRIC PATIENTS: A SINGLE INSTITUTION EXPERIENCE

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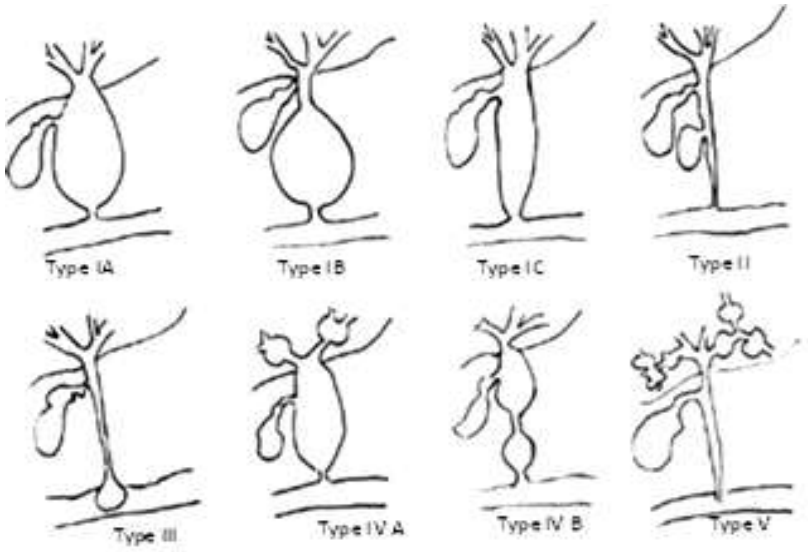
ABSTRACT
Choledochal cyst is an aneurysmal dilatation of the biliary tree involving either extrahepatic or intrahepatic biliary ducts or both, presenting primarily in infants and young children. It is a potentially serious disease with a wide spectrum of presentation. A correct diagnosis and early surgical treatment are important because of the postoperative complications including malignant degeneration. We present our experience with a series of patients with choledochal cyst and discuss its surgical management, with emphasis on reducing the intraoperative and postoperative morbidity. The aim of the study was to present the current modality of surgical treatments i.e; complete excision with Roux-en-Y hepaticojejunostomy and Lilly's procedure, and derive appropriate management recommendations in patients of choledochal cyst. A prospective study conducted in the department of Paediatric Surgery, SMS Medical College, Jaipur between may 2007 to may 2010 is presented. The study included 55 cases of choledochal cyst. Clinical and surgical details of these patients were recorded. Patients were followed up for 3 to 6 years duration. There were 55 patients presenting between the age from new born to 12 years. Jaundice was the most common initial symptom. Eight patients (14.5%) had the triad of jaundice, abdominal pain and palpable mass. Fifty three (96.36%) patients were affected by a cystic dilatation type I of Todani classification, rest two were of different types. Operative procedure was carried out in 52 cases. Lilly's procedure was performed in 25(48%) patients, while 27(52%) underwent complete excision of the extrahepatic duct, cholecystectomy and Roux-en-Y hepaticojejunostomy. Early post-operative complications (<3 months postoperatively) were observed in 5 (9.6%) cases. Late complications (3 months -3 years postoperatively) were seen in 10 (19.2%) patients. No patient had common bile duct adenocarcinoma. There was no operative mortality. Out of the 52 operated patients, 30 patients have been lost to follow up but the remainder, 22 patients are well after 3-6 years. Although the definitive treatment of Choledochal cyst is complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepatojejunostomy, but removal of adherent cyst wall to portal vein had no added benefit, rather it increased the morbidity in terms of intraoperative blood loss and postoperative complications. We believe and lay equal emphasis towards Lilly's procedure in cases with cyst densely adherent to the portal vein secondary to long-standing inflammation because of low surgical morbidity and mortality associated with it. Moreover malignant transformation did not occur in any patient. Follow-up is essential to detect complications, such as cholangitis, anastomotic stricture, intrahepatic cholelithiasis and cholangiocarcinoma.

Keywords: *Choledochal Cyst, Pediatric, Roux-en-Y Hepaticojejunostomy, Lilly's Procedure*

INTRODUCTION
Choledochal cyst is a rare congenital abnormality of the biliary tract and it accounts for approximately 1% of all benign biliary diseases (Saxena *et al.*, 1988). It is defined as a aneurysmal dilatation of the biliary tree involving either extrahepatic or intrahepatic biliary ducts or both, occurring in approximately 1 in 100,000-150,000 live births in the western population (O'Neill *et al.*, 1987; Gigot *et al.*, 1996). 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. Incidence of Choledochal cysts is 1 in 13,500 live births in the United States and 1 in 15,000 in Australia (Gigot *et al.*, 1996). Most of the reported cases in the world come from East Asia. It is common in Japan, with an incidence of 1:1000 (Uribarrena *et al.*, 2008). The first reported case of choledochal cyst was described by Vater & Ezler in 1723 (Skapinker, 1956). Initial classification by Alanso-Lej *et al.*, classified choledochal cysts into three main types, type I–III on

clinical and anatomic findings in 1959 (Alonso *et al.*, 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 (Todani *et al.*, 1977; Miyano *et al.*, 1996). 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.



Type I-Dilatation of hepatic and common bile ducts
IA- Dilatation of extrahepatic bile ducts (entire)
IB- Dilatation of extrahepatic bile ducts (focal segment)
IC- Dilatation of the CBD portion of extrahepatic bile ducts
Type II-Diverticulum of the common bile ducts
Type III-Intraduodenal common bile ducts dilatation (choledochocele)
Type IVA-Intrahepatic and extrahepatic bile ducts dilatation
Type IVB-Multiple extrahepatic cysts
Type V- Intrahepatic bile ducts dilatation (as in Caroli's Disease)

Figure 1: Diagrammatic representation of Todani modified Alonso-Lej classification system of choledochal cysts

At present, the basic principle of management of choledochal cyst is excision of the entire dilated extrahepatic bile duct without damaging the adjacent vital structures such as the portal vein, the hepatic artery, the pancreas and the duodenum by meticulous dissection. The different types of anatomical variants of the hepatic arteries and the bile duct also increase the difficulty of surgery and increase the morbidity. We present our experience with a series of patients with choledochal cyst and discuss its surgical management, with emphasis on reducing the intraoperative and postoperative morbidity.

Aims

To present the current modality of surgical treatments i.e; complete excision with Roux-en-Y hepaticojejunostomy and Lilly's procedure, and derive appropriate management recommendations in patients of choledochal cyst.

MATERIALS AND METHODS

Patients and Methods

Patients affected with choledochal cysts, who were admitted in the Department of Paediatric Surgery, SMS Medical College, Jaipur between May 2007 to May 2010 were studied prospectively. The study included 55 cases of choledochal cyst. We studied demographic data (age, sex), clinical data, presenting

symptoms, medical comorbidities, and diagnostic modalities including hepatobiliary US (ultrasound), CT scan (Computed Tomography) of abdomen. Surgical records including surgical strategy, complications and morbidity and mortality were recorded. Patient follow-up after hospital discharge was done in the outpatient service. During follow-up, laboratory studies were conducted (liver function tests) and imaging (especially US). Patients were followed up for 3 to 6 years duration.

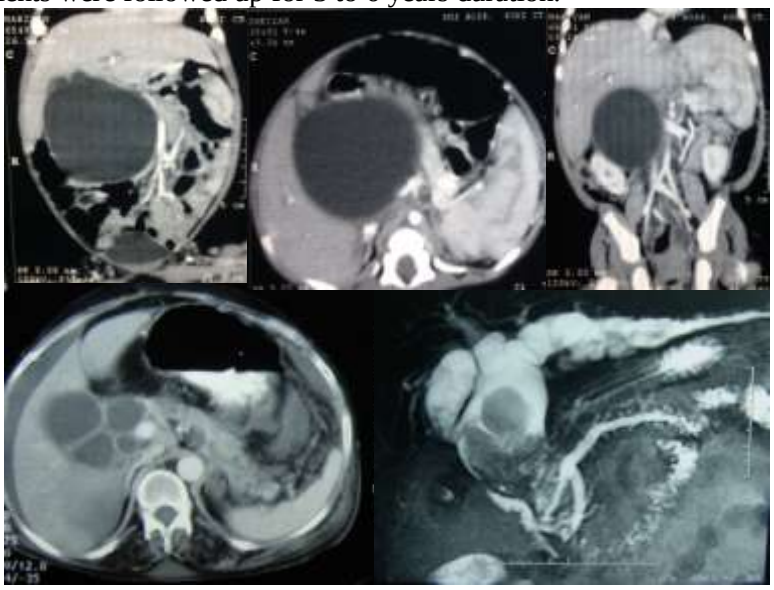


Figure 2: Top photos: CT-scan images of choledochal cyst Type IA. Bottom photos: MRCP- images of choledochal cyst Type IV A

RESULTS

The results of the study are summarized in Table-1 to Table-5. There were 55 patients, 30 (54.5%) boys and 25 (45.5%) girls, with male-to-female ratio of 1.2:1. The age of patients ranged from new born to 12 years. Maximum number of patients, 17 (30.9%) were between 5-7 years of age, while 43 (78.2%) patients presented younger than 7 years of age. Jaundice was the most common initial symptom, present in 36(65.5%) patients. Eight patients (14.5%) had triad of jaundice, abdominal pain and palpable mass.

Table 1: Frequency of choledochal cyst types according to Todani's classification

Type of cyst	Number of patients (%age)
IA	53(96.36)
II	01(1.8)
IVA	01(1.8)
Total	55(100)

Table 2: Presentation of choledochal cyst

Mode of presentation	Number (%)
Jaundice	36(65.5%)
Abdominal pain	35 (63.6%)
Dark urine	11 (20%)
Acholic feces	22 (40%)
Abdominal mass	17 (30.9%)
Cholangitis	08 (14.5%)
Triad	08 (14.5%)
Antenatal ultrasonography	0
Portal hypertension	01(1.8)
Pancreatitis	01(1.8)
ASD, VSD, cleft lip and cleft palate (Died)	01(1.8)

Table 3: Distribution of patients and the surgical procedure, according to the age at diagnosis

Age (years)	Number of patients (%age)	Male (Boys)	Females (Girls)	Male Lilly's	Female Lilly's	Male Total excision	Female Total excision
Neonate	1(1.8)	0	1	0	0	0	0
<1 year	7(12.7)	6	1	1	1	5	0
1-3	10(18.2)	5	5	4	2	1	3
3-5	8(14.5)	4	4	3	1	1	3
5-7	17(30.9)	8	9	4	6	3	3
>7	12(21.8)	7	5	2	1	4	4
Total	55(100)	30 (54.5)	25 (45.5)	14	11	14	13
		55(100%)		25(48%)		27(52%)	
				52(100%)			

Table 4: Early post-operative complications (<3 months postoperatively)

Early complications	**N**
Anastomotic biliary fistula	1
Post-operative Intususception	1
Burst abdomen	1
Cholangitis	2
Total	5 (9.6%)

Table 5: Late complications (3 months -3 years postoperatively)

Late complications	**n**
Biochemical liver dysfunction	4
Recurrent abdominal pain	2
Residual lithiasis in the hepatic duct	1
Common bile duct adenocarcinoma	0
Chronic pancreatitis	1
Portal hypertension	1
Internal hernias of the small bowel	1
Total	10 (19.2%)

Prenatal diagnosis was not performed in any of the 55 patients. Forty eight (87.2%) patients were diagnosed by sonography of the abdomen. CT scan of the abdomen was carried out in 54(98.2%) patients in order to corroborate the diagnosis. Endoscopic retrograde cholangiopancreatography was carried out in one patient because of recurrent multiple episode of biliary colic, gallstones and dilatation of the biliary tract. It indicated an abnormal common hepatic duct and ERCP stenting was performed. A long pancreaticobiliary common channel was, however not demonstrated in any of our cases, as the radiologists probably did not look specifically for this anomaly.

Operative procedure was carried out in 52 cases. Age of patients ranged from 5 months to 12 years. 27(52%) patients with type I underwent excision of the extrahepatic duct, cholecystectomy and Roux-en-Y hepaticojejunostomy, including one patient with type II (because of the broad base of the cyst), and one patient with type IV cyst. Lilly's procedure was performed in 25(48%) patients, with cyst being adherent to the portal vein. Concomitant hepatic biopsies were carried out in 4(7.7%) patients.

Post-operative complications were observed in 15 (28.8%) cases. The early post-operative complications (<3 months postoperatively) were observed in 5 (9.6%) cases. Anastomotic biliary fistula in one case was managed conservatively with reinsertion of sub-hepatic drain. Post-operative Intususception in one patient was managed by per-operative manual reduction. Two (3.6%) patients presented with cholangitis, managed medically (antibiotics and ursodeoxycholic acid). Burst abdomen seen in one case was treated with tension suturing and it was the only complication seen in patients undergoing Lilly's procedure.

Late complications (3 months -3 years postoperatively) were observed in 10 (19.2%) patients. They were biochemical liver dysfunction in 4 patients, managed medically. Recurrent abdominal pain was seen in 2 cases. Residual lithiasis in the hepatic duct present in 1 case was managed with redo-hepaticojejunostomy. Portal hypertension and chronic pancreatitis was present in 1case each. Internal hernias of the small bowel seen in one female patient 3 years after the procedure and was managed surgically successfully. All the late complications were seen in the patients who underwent total excision, while none in patients with Lilly's procedure.

There were no patient deaths during the first 30 days postoperatively, so there was no operative mortality. None of the patient had malignant transformation of the residual cyst wall. Out of the 52 patients who were operated, 30 patients have been lost to follow up but remainder, 22 patients are well after 3-6 years.

DISCUSSION

Choledochal cysts are usually diagnosed in childhood, with 60% diagnosed before the age of 10 years, while only 25% are detected in adult life (Kim *et al.,* 1995; Liu *et al.,* 2002). They also have an unexplained female: male preponderance, commonly reported as 4:1 to 3:1 (Liu *et al.,* 2002). It is commonly reported as 1.5: 1 in the paediatric population (Hung *et al.,* 2011). In our study, male preponderance was present, in contrast to the reported series in the literature. We believe that the gender discrimination prevalent in our society and difference in rearing of the female child is the cause of under-reporting of the female cases in this part of the country.

Aetiology

The aetiology of Choledochal cyst remains speculative, however the pathogenesis is multifactorial in origin (Singham *et al.,* 2007). Choledochal cyst may be the result of an embryonic malformation causing weakness of the wall of the bile duct (Linder and Green, 1964). This weakness is coupled with a distal obstruction to produce the typical cystic anomaly (Alonso *et al.,* 1959). Antenatal ultrasound has documented the presence of choledochal cyst in utero, making it a congenital disease in nature. The Babbitt's theory, Anomalous pancreaticobiliary duct junction (APBDJ) remains the most popular and accepted. It holds true only for children and young adolescents (90-100% of patients with choledochal cysts) (Babbitt's, 1969; Miyano *et al.,* 1997). Pancreaticobiliary malunion may occur without choledochal dilatation, and this has been termed a forme fruste choledochal cyst (Miyano *et al.,* 1997). Other etiologies proposed are abnormal function (spasm / dysautonomia) of the sphincter of Oddi, inadequate autonomic innervations with abnormally few ganglion cells in the narrow portion of the common bile duct, duodenal or biliary duplication cysts, perinatal infectious cholangitis, viral infections, most likely reovirus (Schweizer *et al.,* 1993; Kusunoki *et al.,* 1988; Kagiyama *et al.,* 1987; Landing, 1974; Tyler *et al.,* 1998).

Presentation

Infants who have been diagnosed with a choledochal cyst prenatally do not ordinarily become jaundiced until 1 to 3 weeks after birth. Presentation is usually at the age of 1 to 3 months in the infantile form, with jaundice and acholic stools. Clinical picture is indistinguishable from that of Biliary atresia. Palpable mass in the right upper abdomen, with or without hepatomegaly is seen in late infancy (Douglas, 1852; Watanatittan *et al.,* 1998; Stringer *et al.,* 1995; Samuel and Spitz, 1996). In the so-called adult form of choledochal cyst, clinical manifestations generally become evident after the age of 2 years. The symptoms are recurrent colicky pain, in the epigastric region and right upper quadrant with radiation (oftenly) to the right scapula followed by fever. This is due to intermittent biliary obstruction. In some patients mild form of jaundice is seen, which may be a pointer towards cholangitis. Other features are palpable right upper quadrant mass, and recurrent bouts of pancreatitis (Douglas, 1852; Watanatittan *et al.,* 1998; Stringer *et al.,* 1995; Samuel and Spitz, 1996). Patients with choledochoceles (type III) are not usually diagnosed until they are at least 5 years of age because the characteristic clinical symptom is abdominal pain of an intermittent nature that is not specific (Kagiyama *et al.,* 1987).

The classic characteristic triad of abdominal pain, jaundice and right hypochondrial mass occurs mostly in childhood (0%-17%) and approximately 7 times more frequently in children than in adults (Douglas,

1852; Watanatittan *et al.,* 1998; Stringer *et al.,* 1995; Samuel and Spitz, 1996). In our series eight patients (14.5%) had the triad of choledochal cyst. Eighty five % of children have at least 2 features of the triad at presentation, compared with only 25% of adults (Lipsett *et al.,* 1994). Abdominal pain is the most common symptom in adults, and may present with severe complications. If cholangitis continues for a long period of time, severe jaundice develops, and may result in septicaemia. Associated anomalies include annular pancreas, duodenal atresia, malrotation, renal agenesis, polycystic kidneys, thoracic hemivertebrae (Dudin *et al.,* 1995).

Complications

Pancreatitis is a potentially serious complication, which is likely due to Anomalous pancreaticobiliary duct junction (APBDJ) leading to the activation of pancreatic enzymes by bile reflux (Swisher *et al.,* 1994). ninety% of the patients with choledochal cysts having a diameter equal to, or greater than, 5 cm developed pancreatitis as compared to only 9% of patients having a choledochal cyst less than 5 cm. The incidence of pancreatitis is higher in children as compared to adults. Pancreatitis is associated with all types of choledochal cysts and is not related to either gender or race (Swisher *et al.,* 1994). Other common complication is cholangitis (Samuel and Spitz, 1996; Nambirajan *et al.,* 2000). Biliary Cirrhosis has been found to occur in 10% to 50% of cases and it's the main mechanisms responsible for the development of Portal hypertension (PHT) in children, although it's a rare complication associated with choledochal cyst (Swisher *et al.,* 1994; Nambirajan *et al.,* 2000; Saluja *et al.,* 2011; Yeong *et al.,* 1982). There is some evidence that even in patients with biliary cirrhosis and portal hypertension, disease regression is possible after surgical treatment (Yeong *et al.,* 1982). One of the rare complications of choledochal cyst is biliary peritonitis from cyst rupture. External bile drainage would be safer in emergency condition, especially when patients present late with sepsis and comorbidities (Swisher *et al.,* 1994).

Choledochal cyst is a premalignant state and malignant change should be suspected in any choledochal cyst appearing after infancy, although in pediatric cases metaplasia and dysplasia are uncommon (Iwai *et al.,* 1990; Rattner *et al.,* 1983). The presence of cholangiocarcinoma in the cyst is reported in <1% in pediatric cases. The overall risk is 10%–15%, and increases with age (Bismuth and Krissat, 1999). The risk with an unresected choledochal cyst remains at 20-30% (Ong *et al.,* 2013). Risk of cholangiocarcinoma in cysts is much higher in patients with APBDJ (32%) than in those without (0%) (Song *et al.,* 1999). Pancreatic juice reflux secondary to APBDJ, chronic inflammation, bile stasis, possible development of carcinogens and decrease of mucosal glands in the bile duct are the predisposing factor (Song *et al.,* 1999). The most frequent site of carcinoma development is the cyst wall, although it may be at any height of the biliary tree (Fieber and Nance, 1997). Cancers are often first diagnosed at laparotomy, and can already be unresectable. Adenocarcinoma is the most common type and the usual site is extrahepatic bile duct (Bismuth and Krissat, 1999).

Investigations

Ultrasound is an excellent first-line investigation with a specificity of 97% in children (Hung *et al.,* 2011; Matsubara *et al.,* 1997). A dilated cystic lesion which communicates with the bile duct, separate from the gall bladder is seen on ultrasound. MRCP is considered "gold standard" in the imaging of choledochal cysts and defining APBDJ (Kim *et al.,* 2000). It is superior CT, but in our resourse limited set up, where facilities for sedating and carrying out the investigation in smaller children compounded with the non availability of trained anaesthetist, makes it an extremely difficult endeavour. We consider CT with contrast as investigation of choice in smaller children. Radionucleotide Scintigraphy is safe and has 100% accuracy to distinguish it from biliary atresia (Rosenfield *et al.,* 1975). Endoscopic Retrograde Cholangiopancreatography (ERCP) is an invasive technique and has potential complications and there is a lack of cooperation from pediatric patients. It should be reserved for adult patients. Also MRCP has been shown to be just as good as ERCP as far the diagnosis of choledochal cyst (Kim *et al.,* 2000).

Surgical Management

Preoperative optimisation is important (Samuel and Spitz, 1996; Nambirajan *et al.,* 2000). Surgery should be performed early after diagnosis, even in asymptomatic prenatally diagnosed neonates (within the first

1–3 months of age), to prevent complications, particularly liver fibrosis in neonates. Cyst-enterostomy internal drainage procedure have a historical value due to unacceptably high rate of late complications (Todani *et al.,* 1977). This is secondary to the fact that the anastomosis should be performed as mucosa to mucosa, whereas the cyst does not have normal mucosa (Todani *et al.,* 1977; Scudamore *et al.,* 1994).

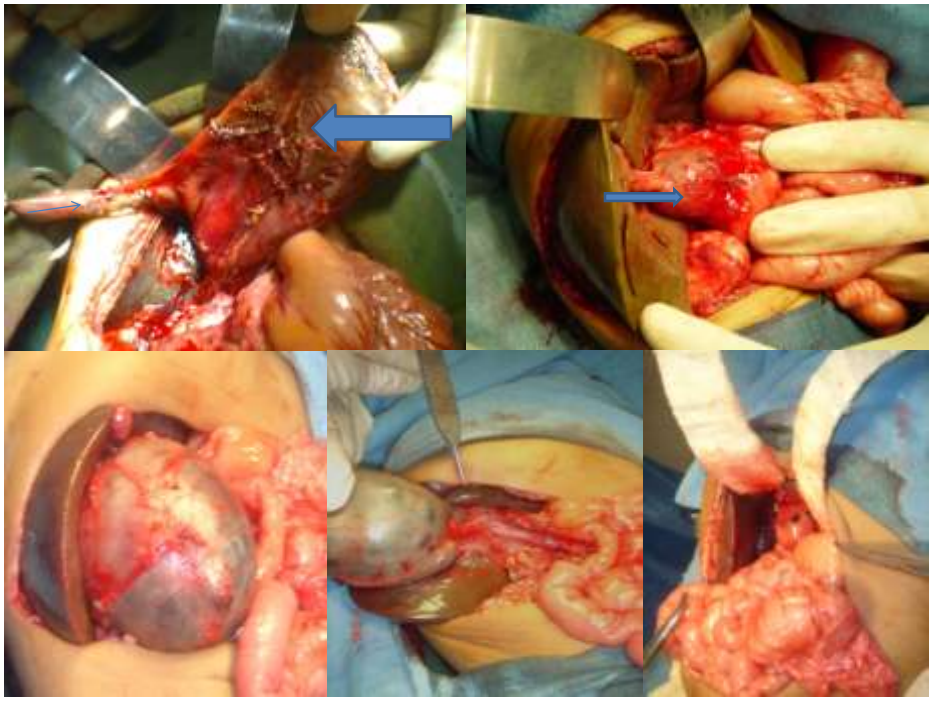


Figure 3: Various peroperative pictures of type IA choledochal cyst (blue arrow), along with the gall bladder (blue line), On bottom right: Peroperative photograph showing completion of Roux-en-y hepatico-jejunostomy

The surgical management of choice for Type I, Type IVB and extrahepatic component of Type IVA is complete excision of the involved portion of the extrahepatic bile duct from the hepatic hilum to the pancreaticobiliary duct junction with construction of a biliary-enteric anastomosis by a Roux-en-Y hepaticojejunostomy to restore continuity with the gastrointestinal tract (Scudamore *et al.,* 1994). In Type II complete excision of the dilated diverticulum, the resultant defect in the common bile duct is closed over a T-tube. Therapeutic choice depends on the size of the cyst in Type III. Choledochoceles measuring 3 cm or less can be treated effectively with endoscopic sphincterotomy, whereas lesions larger than 3 cm (which typically produce some degree of duodenal obstruction) are excised surgically. A longitudinal duodenotomy permits complete exposure of an intraduodenal choledochocele. Once the choledochocele is exposed, it should be unroofed, and then the mucosa is reapproximated with multiple interrupted absorbable sutures. If the pancreatic duct enters the choledochocele, its reimplantation into the duodenum may be required following excision of the cyst (Scudamore *et al.,* 1994).

Intrahepatic dilatation of type IVA cyst in children do not adversely affect the postoperative outcome after conventional surgical repair. It has been suggested that intrahepatic dilatation is related to sustained increase of intrabiliary pressure rather than any intrinsic intrahepatic cystic malformation and that effective surgery invariably reduces measured intrahepatic dilatation towards normal values (Hill *et al.,* 2011). Type IVA intrahepatic ductal disease resection of the affected hepatic segment or lobe is indicated in hepatolithiasis, intrahepatic ductal strictures, and hepatic abscesses. In localized intrahepatic ductal disease relevant partial hepatectomy may be performed due to risk of malignancy (Hill *et al.,* 2011).

In Type V hepatic lobectomy for disease limited to one hepatic lobe and liver transplantation for bilobar disease, liver failure, biliary cirrhosis, and portal hypertension are treatment options (Scudamore *et al.*, 1994). Cholecystectomy with cystic duct ligation near the common bile duct is curative in Type VI.

Cyst is transected at the bifurcation or more proximally if it extends into the individual hepatic ducts. If the Choledochal cyst extends distally into the pancreas, the mucosa of the intrapancreatic portion of the cyst should be stripped away prior to closure at the point of distal transaction. Theoretically, this removes the risk of malignant transformation in that segment of the duct. After excision of the cyst, the intrahepatic ducts should be probed and lavaged with saline to rid ductal system of sludge and possible stones. Additionally, on occasion, obstruction may be found in proximal biliary system, which can be dilatated. Therefore, intraoperative cholangiography before cyst excision is a valuable investigation. Excised cyst should be examined grossly for malignancy and should be sent for frozen section.

Patients with recurrent bouts of cholangitis, pericystic inflammation, the cyst adheres densely to the portal vein and adjacent vascular structures. In these cases periductal or pericystic dissection with full-thickness excision of the cyst is unsafe owing to the dreaded complication of portal venous injury, in particular. Excision is accomplished by removing only the less adherent portion, the posterior wall is dissected using a plane between the inner (mucosa) and outer layers of the cyst (serosal surface) or mucosa of the cyst wall is obliterated by curettage or cautery. Hence, the portion of the cyst wall and bile duct that is adherent to the portal vein and hepatic artery is left undisturbed. This is Lilly's procedure, also known as Lilly's mucosectomy technique (Lilly, 1979; Lee *et al.*, 2009). The mucosal lining of the retained cyst wall should be ablated by diathermy, as 57% of the cholangiocarcinomas in a choledochal cyst arises from the posterior wall of the cyst (Flanigan, 1977). The procedure was performed in 25(48%) cases in our study owing to the same indications proposed by Lilly, while it was necessary in about 10% of the cases in one series (Lee *et al.*, 2009). Although it seems that the percentage of this procedure was large, but we believe that, intra and postoperative complications in our study were quite less in patients undergoing Lilly's procedure.

Postoperative Complications

Postoperative morbidity is 15-30%, mainly due to surgical wound infection, cholangitis and leakage of the anastomosis, which in most cases are managed conservatively (Palanivelu *et al.*, 2008; Visser *et al.*, 2004). In our study, there was no postoperative haematoma or subhepatic collection, as seen in anoter study (Lee *et al.*, 2009). In our study total postoperative complications were seen in 15 (28.8%) patients.

Postoperative anastomotic strictures are associated with intrahepatic cholelithiasis and cholangitis. It is reduced by performing a higher anastomosis, i.e; hepaticojejunostomy rather than choledochojejunostomy (Todani *et al.*, 1998). Reoperation for recurrent cholangitis secondary to stenosis has been reported to be from 13-69% (Chaudhari *et al.*, 1997). A wide anastomosis aims to prevent persistent cholestasis which may be a high-risk factor for carcinogenesis (Chaudhari *et al.*, 1997). Intrahepatic stones/ choledocholithiasis are a particular problem in cases of type IV disease with residual intrahepatic cysts. Choledochoscopy, intraoperative cholangiography with ductal probing and washout of debris reduces postoperative complications of stones retained in the biliary radicals. Intracystic lithiasis is frequently associated with tumour, and can give similar radiological appearances (Dayton *et al.*, 1983). Intestinal obstruction due to adhesions and internal hernias of the small bowel are a recognised complication of roux-en-y bypass procedures (Ong *et al.*, 2013). Postoperative pancreatitis with or without protein plug is rare after cyst excision and internal drainage. Rarely, patients may develop pancreatitis after cyst excision and internal drainage. The morphology of pancreatic duct and ductal dilatation, possibly caused by stasis of pancreatic juice, may be the etiological factor (Swisher *et al.*, 1994).

The risk of post-excision malignancy increases with age, rising from 0.7% in patients with cysts operated on at below 10 years of age, to 6.6% when operated between 11- 20 years, and 14.3% if first treated at > 20 years old (Voyles *et al.*, 1983). It is more common in cysts of Alonso-Lej Types I, IV, and V (Dayton *et al.*, 1983). It is postulated that it can occur in residual intrapancreatic portion of the choledochal cyst, in cases with intrapancreatic extension, and in most of the cases, the cause may be attributed to incomplete resection (Watanabe *et al.*, 1999). Its excision is recommended for its prevention. Postoperative stenosis

and repeated cholangitis lead to bile stasis and intrahepatic lithiasis which may be a high-risk factor for carcinogenesis (Ando *et al.,* 1996). Malignancy has been reported to arise in the intrahepatic cysts, intrahepatic bile duct or residual common bile duct following resection of the cyst (Watanabe *et al.,* 1999; Ando *et al.,* 1996; Ono *et al.,* 2008). In our series out of 52 cases, including 25(48%) who underwent Lilly's procedure, none of the patient had malignant transformation of the residual cyst wall and same holds good for 75 patients, including 10 % cases who underwent Lilly's procedure in one large series (Lee *et al.,* 2009). It is believed that duration of chronic inflammation is expected to be shorter in children and the occurrence of malignancy in children is less as compared to adults (Bismuth and Krissat, 1999; Ong *et al.,* 2013; Song *et al.,* 1999).

Postoperative Follow-Up

To protect against the cholangitis, we keep the patients on low doses of antibiotics for approximately 6 weeks postoperatively. During this period, patient is rigorously assessed clinically for any features of cholangitis. The potential for this complication decreases after this period. Three monthly follow-up for the first year and annually thereafter is recommended. Liver function studies and serum amylase levels are carried at each visit. Annual ultrasound of the liver and pancreas is done, particularly in those having intrahepatic ductal dilatation preoperatively. It is an extremely useful investigation for identifying the postoperative complications.

Conclusion

Choledochal cysts present non-specifically rather than classically. A high index of suspicion will avoid a delay in diagnosis, which may result in a variety of undesirable sequelae. Although the definitive treatment of Choledochal cyst is complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepatojejunostomy, but removal of adherent cyst wall to portal vein had no added benefit, rather it increased the morbidity in terms of intraoperative blood loss and postoperative complications. We believe and lay equal emphasis towards Lilly's procedure in cases with cyst densely adherent to the portal vein secondary to long-standing inflammation because of low surgical morbidity and mortality associated with it. Moreover we didn't have any case of cholangiocarcinoma in our study. Follow-up is essential to detect development complications, such as cholangitis, anastomotic stricture, intrahepatic cholelithiasis and cholangiocarcinoma.

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