Conventional and unconventional surgical modalities for choledochal cyst: long-term follow-up

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Background **This study presents our experience of various modes of presentation, surgical methods applied (both conventional and unconventional), and their outcome on patients with choledochal cyst.**

Method **This is a retrospective study in which files of all patients with choledochal cyst over a period of 10 years (1997–2007) were studied. Data collected from files included age, sex, modes of presentation, various surgical techniques, outcome, and follow-up.**

Results **The total number of patients was 35. The most common mode of presentation was recurrent abdominal pain in 20 (57%) patients. It was followed by jaundice in 16 (45%) patients, fever, nausea, and vomiting in 10 (28.5%) patients, and other symptoms. Roux-en-Y hepaticojejunostomy was performed in 26 (74%) patients, hepaticoduodenostomy in five (14%) patients, external T-tube drainage in three (8%) patients, and cystoduodenostomy in two (5%) patients. Two patients expired in this series. The follow-up loss was 20%. Three patients who suffered from cholangitis were managed conservatively. No patient has shown cirrhosis and cholangiocarcinoma till now.**

Conclusion **Choledochal cyst is an important entity in an Indian setup. The presentation has a wide spectrum; therefore, different types of surgical intervention would play a significant role in various situations.**
Ann Pediatr Surg **7:16–18 © 2011 Annals of Pediatric Surgery**

Annals of Pediatric Surgery 2011, **7**:16–18

Keywords: choledochal cyst, follow-up of choledochal cyst, surgical techniques

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Received 15 October 2010 **Accepted** 17 November 2010

Introduction

Choledochal cyst is a congenital biliary tract anomaly that may involve extrahepatic, intrahepatic, or even both biliary radicals [1]. The disease is characterized by various types of abnormal biliary tract dilations, which forms the basis of its classification, initially given by Alonso Lej *et al.* [2] and subsequently modified by Todani *et al.* [3]. This disease is more prevalent in Asian countries [1]. Females, according to the literature, show eight times more predominance of the disease than males [1]. Although prenatal ultrasonography (USG) of fetal choledochal cyst has been reported by a number of investigators, the majority of these patients present during childhood, which makes this disease crucial to be diagnosed and managed promptly [1].

This study shares our experience of various modes of presentation, the different surgical modalities (both conventional and unconventional) applied, and their effect on the overall survival of these patients.

Material and methods

In this retrospective study, all patients with choledochal cyst admitted from January 1997 to December 2007 in the Department of Pediatric Surgery (CSM Medical University, India) were included. The patients were analyzed on the basis of age at presentation, sex, mode of presentation, diagnostic modalities used, types of surgical intervention applied, and their outcome on follow-up.

All patients were evaluated clinically, supplemented by radiological interventions like abdominal USG, abdominal computerized tomography, magnetic resonance cholangiopancreatography, and hydroxy iminodiacetic acid scan. They were followed in the same manner on every visit.

The various procedures used in our series were Roux-en-Y hepaticojejunostomy, hepaticoduodenostomy, cystoduodenostomy, and external T-tube drainage. Cystoduodenostomy was performed in those patients who had evidence of cirrhotic changes of the liver at the time of surgery. If the common hepatic duct was long enough to be anastomosed with the duodenum, hepaticoduodenostomy was performed. The external T-tube drainage was performed in patients with a ruptured choledochal cyst. In the postoperative period, the patients were kept nil per os for 5 to 7 days. After discharge, the patients were called in the outpatient department after 15 days. Liver functions (which included serum direct, indirect, and total bilirubin, serum glutamic oxaloacetic transaminase, serum glutamic pyruvic transaminase, and alkaline phosphatase) were assessed at that time and after 2 months. If there was raised bilirubin in the postoperative period persistently or a rise in bilirubin levels after coming to near normal values without the evidence of cholangitis, such as coexisting fever, a hydroxy iminodiacetic acid scan was taken. If the patients had no problems they were called after 6 months or they themselves consulted for any problem, if it arose. Cholangitis was suspected on clinical evidence of fever and jaundice, supplemented by the liver functions. It was treated by antibiotics, which included injections of ceftriaxone (50 mg/kg intravenously every 12 h), amikacin (7.5 mg/kg

1687-4137 © 2011 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000393093.1853.99

intravenously every 12 h) for 7–10 days, and mertonidazole (5 mg/kg intravenously every 8 h) for 5 days. The patients responded to this treatment.

Results

The duration of this study was 10 years. The total number (*n*) of patients was 35. Choledochal cyst was found more commonly in the patients aged between 1 and 5 years ($n = 23$), followed by those aged between 6 and 10 years ($n = 9$), with only three cases ($n = 3$) recorded in patients aged less than 6 months. The mean age was 4.39 ± 2.58 years. The male to female ratio was approximately 2 : 1.

Clinically, recurrent abdominal pain was present in 20 (57.14%) patients. Other complaints that were noticed were jaundice, fever, nausea and vomiting, palpable lump in abdomen, peritonitis, and ascites (Table 1). Todani types 1, 2, and 4a were encountered in our series. Of these, type 1 was found to have the maximum occurrence in our series (Table 2).

Roux-en-Y hepaticojejunostomy was the most common operation that was performed. It was followed by hepaticoduodenostomy, cystoduodenostomy, and external T-tube drainage (Table 3).

The complications noticed were cholangitis in three patients, two of them in Roux-en-Y hepaticojejunostomy and one in cystoduodenosyomy. All of these were managed conservatively. Another single case of type 1 choledochal cyst, after undergoing Roux-en-Y hepaticojejunostomy, had presented in emergency, 1 week after discharge, with acute intestinal obstruction, for which exploratory laparotomy was performed and was found to have terminal ileal gangrene. The distal ileum was exteriorized by ileostomy, which was later repaired with resection and anastomosis. The etiology for such an event could not be found, as the histopathology was reported to be inconclusive.

Table 1 Modes of presentation of patients with choledochal cyst

Signs and symptoms[a]	Number of patients (*n*, %)
Recurrent abdominal pain	20 (57.14)
Jaundice	16 (45.7)
Fever, nausea, vomiting	10 (28.57)
Lump in abdomen	5 (14.28)
Peritonitis	3 (8.57)
Ascitis	2 (5.71)

[a]One or more sign or symptoms may be present in a single patient.

Table 2 Types of choledochal cysts

Todani types	Number of patients (*n*, %)
Type 1[a]	28 (80)
Type 2	4 (11.42)
Type 3	0
Type 4a	3 (8.57)
Type 4b	0
Type 5	0

[a]The most common of all types of choledochal cyst.

Table 3 Surgical modalities used for the treatment of choledochal cyst

Type of surgery	Number of patients (*n*, %)
Roux-en-Y hepaticojejunostomy[a]	26 (74.28)
Hepaticoduodenostomy	5 (14.28)
Cystoduodenostomy	2 (5.71)
External T-tube drainage	3 (8.57)

[a]The most common operation performed for choledochal cyst.

Follow-up loss was approximately 20%. Two patients (5.71%) expired in our series. The mean follow-up period was 2.83 ± 1.78 years.

Discussion

Vater [1] had first described a choledochal cyst with studies of normal and abnormal anatomy of the biliary tree. Later, Alonso Lej *et al.* [2] revolutionized this entity by classifying it into three types, which was subsequently modified by Todani *et al.* [3].

In our study, the most common age group was 1–5 years, which corresponds with other studies [4,5]. Males predominated in our series, whereas in other studies females had accounted for 80% of all cases [1,4,5]. The reason for this observation could not be ascertained. It may be due to more care given to male children in parts of northern India. Although pain was the most common presentation in our series, jaundice has prevailed in other studies [1,3]. The classical triad of pain, jaundice, and lump is an uncommon entity, and was found in three patients, which corresponds with other studies [3,6,7]. Biliary peritonitis due to cyst rupture, although a rare entity in some studies [8,9], needs emergency treatment. It can be diagnosed by USG-abdomen [9] or di-isopropylphenylcarbamoyl-methylimido diacetic acid scan [8]. Todani type 1 was found in 28 patients, in which cystic and fusiform varieties occurred with equal frequencies in both the sexes and in all age groups, unlike other studies in which cystic varieties predominated [10]. Roux-en-Y hepaticojejunostomy was the most commonly performed operation in our series just as in other studies [1,3,5,7,11–14]. Sometimes, there is laxity of the fascial planes created due to the space occupied by the lump [6]. In these cases, hepaticoduodenostomy can be easily performed, as the duodenum can be mobilized. There can be concerns of cholangitis in these patients but it has not been noticed till now. Biliary peritonitis due to ruptured choledochal cyst can be diagnosed on an USG-abdomen scan on a basis of high suspicion. These patients were initially managed by external T-tube drainage and later repaired with Roux-en-Y hepaticojejunostomy as a definitive procedure. We did not perform Percutaneous Transhepatic Cyst (PTC) drainage, which was used as a bridge procedure [15]. We believe it to be invasive and thus, left it unattempted in our patients. Cystoduodenostomy, which has not been used much in the past, was also performed on two patients similar to other studies [6] that had presented with ascites and on an USG scan were found to have some cirrhotic changes in the liver. Surprisingly, one patient is still being followed and is doing well, whereas the other patient was lost to follow-up. Importantly, there has not been a single follow-up

evidence of cirrhosis or cholangicarcinoma noted till date [13].

We emphasize that, although Roux-en-Y hepaticojejunostomy is the gold standard procedure, there are some patients who would present to us in other ways and make the other procedures significant to provide better management and outcome of choledochal cyst.

Conclusion

To conclude, choledochal cyst is an important entity in an Indian setup. Mostly, it presents in the age group of 1–10 years. The presentation has a wide spectrum; therefore, different types of surgical interventions would play a significant role in various situations.

Follow-up forms an important part to look for and treat possible complications.

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