e-ISSN: 0975-9506, p-ISSN:2961-6093

Available online on www.ijpqa.com

International Journal of Pharmaceutical Quality Assurance 2026; 17(3); 103-108

Original Research Article

An Overview of Surgical Management of Choledochal Cysts in Adults: Prospective Study from a Tertiary Center

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Received: 11-01-2026 / Revised: 15-02-2026 / Accepted: 23 -03-2026
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Conflict of interest: Nil

Abstract

Introduction: Choledochal cysts, rare congenital anomalies of the biliary tract, typically present in childhood, yet in 20% of patients the diagnosis is delayed until adulthood. Presentation in adult-life is often associated with other hepatobiliary pathologies, complicating surgical management. These may remain asymptomatic. Symptomatic patients mostly present with symptoms mimicking calculous biliary tract disease. Diagnosis is made by imaging of the biliary tree, can also be incidental. Surgical management is the definitive treatment, dictated by the cyst type along with the presence of associated hepatobiliary pathology.

Aims: To highlight the clinical features, diagnostic approach, and management of an extremely rare case of a giant choledochal cyst.

Materials and Methods : An observational, prospective study was conducted on 36 patients, meeting the study criteria, attending General Surgery OPD at Medical College and Hospital, Kolkata (June, 2022 - June, 2024) treated with cyst excision with Roux-en-Y Hepaticojejunostomy. Patients were followed up for six months postoperatively.

Results: Majority were female (63.6%), median age 38.5 years. Most presented with right upper quadrant pain and jaundice (54.5%). Commonest was Type IVa (68.8%), abnormal pancreatico-biliary junction:13.8%. Commonest associated pathology was cholelithiasis (54.5%). Post-operatively, median hospital stay was 8 days. Most common complication: Surgical site infection (22.2%). Outcomes: Uneventful post-operative course-56.%, Clavien-Dindo Grade I-25%, Grade II-13%, Grade IVa- 5.6%. The median day of abdominal drain removal-Day 6.5, initiation of enteral nutrition-Day 3, bile leak in 8.3%. Bile: Culture-no growth (58.6%), Amylase-raised (22.7%). HPE: non-malignant in 100%. Follow up: Deranged LFT- 2.8%, incisional hernia: 5.6%.

Conclusion: Choledochal cysts are rare congenital biliary anomalies that require timely diagnosis and appropriate surgical management to prevent complications such as cholangitis, pancreatitis, and malignancy. Early recognition using appropriate imaging modalities allows for definitive surgical planning. Complete excision of the cyst with biliary reconstruction remains the treatment of choice and is associated with good postoperative outcomes. The management of a giant choledochal cyst poses additional surgical challenges due to its size, anatomical distortion, and risk of complications; however, successful surgical excision can be achieved with careful planning and expertise. Overall, surgical management provides favorable outcomes with low morbidity when performed appropriately, emphasizing the importance of early intervention and standardized surgical techniques.

Keywords: Choledochal cyst; Giant choledochal cyst; Biliary cyst; Biliary tract anomalies; Cyst excision; Hepaticojejunostomy; Biliary reconstruction; Pediatric and adult biliary disease; Surgical management; Bile duct anomalies.

DOI: 10.25258/ijpqa.17.3.19

Introduction

Choledochal cysts or bile duct cysts are rare congenital anomalies, which usually present a surgical problem in infancy or childhood. However, in about 20% of patients, the diagnosis is delayed

until adulthood. The presentation and management in adults may differ substantially due to an increased rate of associated hepatobiliary and pancreatic pathologies. Hence, the surgical management of choledochal cysts in adults is considerably complex due to the presence of co-existing pathologies and can also be technically challenging because of the added difficulties of re-operative biliary surgery [1].

Choledochal cysts are classified based on the site, shape, and extent of the cystic anomaly of the biliary ductal system. The first systematic classification of extrahepatic biliary cysts was proposed by Alonso-Lej and colleagues in 1959 [2]. This was later refined by Todani in 1977 by integrating variants of Caroli disease (intrahepatic cysts) with the pre-existing classification, and it remains the most widely accepted system for clinical categorization [3]. The Todani classification system divides choledochal cysts into six major types. Type I biliary cysts are solitary fusiform or saccular extrahepatic cysts and are further subdivided based on the gallbladder and cystic duct location. Type II cyst is a supraduodenal diverticulum of the extrahepatic biliary tree. Type III cysts are intraduodenal diverticulum of the distal common bile duct, also known as a choledochocele. Type IV cysts include both intra- and extrahepatic components and are subdivided into Type IVa and Type IVb. Type V cysts involve only the intrahepatic bile ducts and are known as Carolidisease. Newer entities such as Type Id and Type VI cysts have also been described [4]. Although the exact etiology remains unknown, multiple theories of etiopathogenesis have been proposed. The most widely accepted hypothesis suggests that cystic dilatation of the biliary tree is secondary to an anomalous pancreatobiliary ductal junction. This results in a long common channel that allows reflux of pancreatic enzymes into the biliary system, leading to inflammation and subsequent cystic dilatation[5]. Some studies have suggested a hereditary predisposition, while others propose that oligoganglionosis in the distal neck of the cyst may play a role. Biliary cysts may remain asymptomatic for years and later present in adulthood. They are often incidentally detected during imaging for unrelated conditions. Symptomatic patients typically present with features resembling calculous biliary disease, including intermittent right upper abdominal or epigastric pain, fever, and jaundice. The pain may radiate to the back or intrascapular region and can persist for several hours. Some patients may present with cholangitis, worsening jaundice, or features of sepsis. An abdominal lump is uncommon but, if present, may suggest cyst-related complications or malignancy. Giant choledochal cysts may present with a palpable abdominal mass. Pancreatitis may be seen in a significant proportion of patients [5]. A subset of patients may develop secondary biliary cirrhosis with features of portal hypertension due to chronic biliary obstruction and stasis [6]. Accurate preoperative diagnosis is achieved using imaging modalities such as abdominal ultrasound, computed tomography, and magnetic resonance cholangiopancreatography. Magnetic resonance cholangiopancreatography is a non-invasive and highly effective tool for identifying and classifying the cyst type, while contrast-enhanced computed tomography helps in evaluating associated pathologies [7]. Common associated hepatobiliary conditions include cystolithiasis, gallstones, chronic pancreatitis, cholangiocarcinoma, intrahepatic abscess, and secondary biliary cirrhosis. The surgical management depends on the cyst type and usually involves complete excision of the extrahepatic cyst followed by biliary reconstruction, most commonly via Roux-en-Y hepaticojejunostomy. Management of intrahepatic cysts remains controversial, although liver transplantation is considered the treatment of choice for Carolidisease.

Materials and Methods

Study Type: This was an observational, prospective study.

Study Duration: The study was conducted over a period of two years, from June 2022 to June 2024.

Study Place: The study was conducted at the Department of General Surgery, Medical College and Hospital, Kolkata, West Bengal, India.

Study Population:

The study population included patients with choledochal cysts attending the General Surgery Out-Patient Department of Medical College and Hospital, Kolkata, who met the eligibility criteria and underwent surgical management.

Sample Size: A total of 36 patients were included in the study.

Inclusion Criteria

- Patients aged between 18 and 65 years
- Newly diagnosed cases of choledochal cysts
- Patients giving informed consent
- Patients planned for surgical management at the study centre

Exclusion Criteria

- Patients outside the specified age group
- Patients not willing to give consent
- Patients with previously treated choledochal cysts
- Patients unfit for surgery due to severe systemic illness or contraindications to anesthesia

- Patients lost to follow-up or not completing the study protocol

Results

Table 1: Demographic and Clinical Profile (Sample Size = 36)

Parameter	Number of Patients	Percentage
Median age	38.5 years	—
Female	23	63.60%
Male	13	36.40%
Right upper quadrant pain with mild jaundice	20	54.50%
Right upper quadrant pain only	7	20%
Nonspecific gastrointestinal symptoms	6	17%
Abdominal lump	3	9.00%

Table 2: MRCP Findings and Cyst Types

Parameter	Number of Patients	Percentage
Type Iva	24	68.80%
Type I	8	22.70%
Type II	2	4.50%
Type VI	2	4.50%
Anomalous pancreatobiliary junction	5	13.80%

Table 3: Associated Hepatobiliary Conditions

Condition	Number of Patients	Percentage
Cholelithiasis	20	54.50%
Cystolithiasis	8	22.70%
Cholangitis	10	28%
Recurrent pancreatitis	12	33.30%
Secondary biliary cirrhosis with portal hypertension	4	11.10%

Table 4: Giant Choledochal Cyst Case

Parameter	Details
Number of patients	1
Type	Type IVa
Size	26 × 30 cm

Table 5: Surgical Management

Procedure	Number of Patients	Percentage
Choledochal cyst excision with Roux-en-Y hepaticojejunostomy	36	100%
Additional lateral pancreaticojejunostomy	2	5.55%

Table 6: Postoperative Recovery

Parameter	Median Value
Return of intestinal peristalsis	2.5 days
Initiation of enteral nutrition	3 days
Drain removal	6.5 days
Discharge	8 days

Table 7: Intraoperative Bile Culture and Biochemical Findings

Parameter	Number of Patients	Percentage
No growth	21	58.60%
Escherichia coli	9	24.50%
Klebsiella species	7	18.50%
Raised amylase in bile	8	22.70%
Associated with ABPJ	5	13.80%

Table 8: Histopathology Findings

Parameter	Number of Patients	Percentage
Malignancy	0	0%

Table 9: Postoperative Complications

Complication	Number of Patients	Percentage
Surgical site infection	8	22.20%
Pulmonary complications	6	16.60%
Cholangitis	4	11.10%
Bile leak	3	8.30%
Entero-enteric anastomotic leak	1	2.80%
Re-exploration required	2	5.55%
Readmission within 1 month	3	8.33%

Table 10: Clavien-Dindo Classification of Outcomes

Grade	Number of Patients	Percentage
Uneventful recovery	20	56%
Grade I	9	25%
Grade II	5	13%
Grade IV A	2	5.60%

Table 11: Follow-Up Outcomes at 6 Months

Parameter	Number of Patients	Percentage
Incisional hernia	2	5.60%
Liver function derangement	1	2.80%
Anastomotic stricture (on MRCP)	1	2.80%

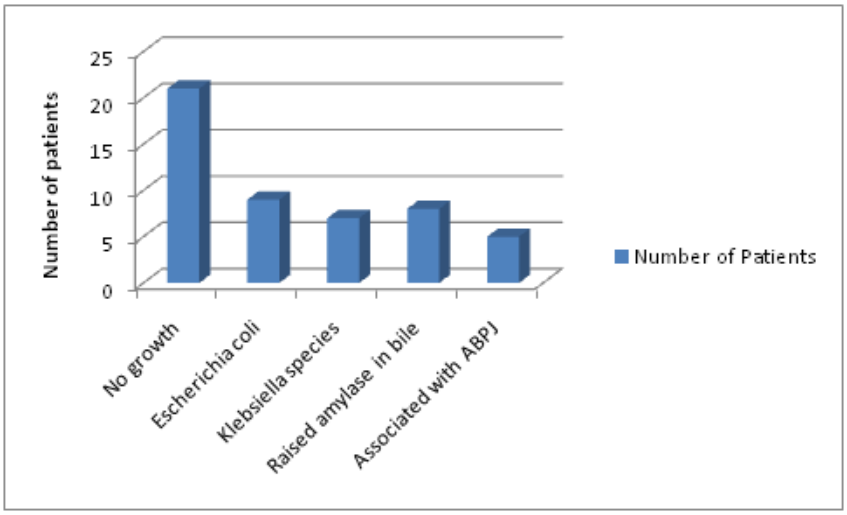

Figure 1: Intraoperative Bile Culture and Biochemical Findings

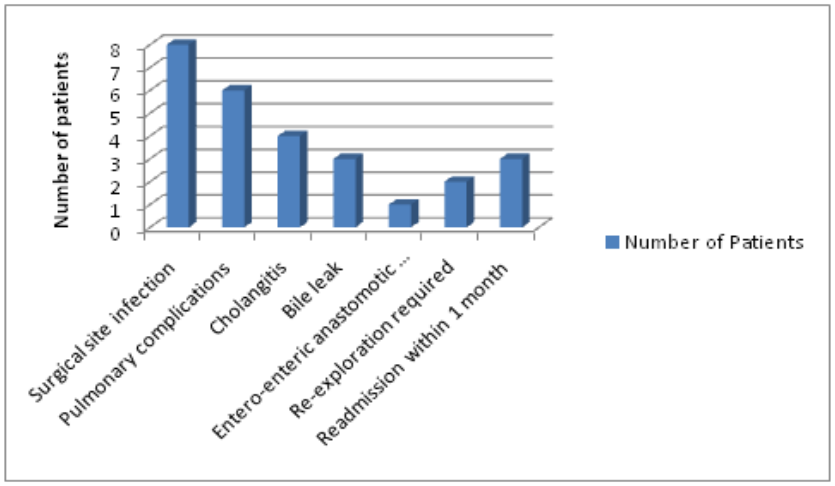

Figure 2: Postoperative Complications

In the present study, out of 36 patients, 23 (63.6%) were females and 13 (36.4%) were males, with a median age of 38.5 years. The most common presenting symptom was right upper quadrant pain with mild jaundice in 20 patients (54.5%), followed by isolated right upper quadrant pain in 7 patients (20%), nonspecific gastrointestinal symptoms in 6 patients (17%), and abdominal lump in 3 patients (9%).

Among 36 patients, Type IVacholedochal cyst was observed in 24 patients (68.8%), followed by Type I in 8 patients (22.7%), Type II in 2 patients (4.5%), and Type VI in 2 patients (4.5%). An anomalous pancreatobiliary junction was present in 5 patients (13.8%).

Out of 36 patients, cholelithiasis was present in 20 patients (54.5%), cystolithiasis in 8 patients (22.7%), cholangitis in 10 patients (28%), recurrent pancreatitis in 12 patients (33.3%), and secondary biliary cirrhosis with portal hypertension in 4 patients (11.1%).

A single patient (1, 2.8%) presented with a giant choledochal cyst measuring 26 × 30 cm, classified as Type Iva.

All 36 patients (100%) underwent choledochal cyst excision with Roux-en-Y hepaticojejunostomy, while 2 patients (5.55%) required additional lateral pancreaticojejunostomy due to associated chronic pancreatitis.

Postoperative recovery parameters showed that the return of intestinal peristalsis occurred at a median of 2.5 days, initiation of enteral nutrition at 3 days, drain removal at 6.5 days, and discharge at a median of 8 days in the study population.

In 36 patients, intraoperative bile culture showed no growth in 21 patients (58.6%), Escherichia coli in 9 patients (24.5%), and Klebsiella species in 7 patients (18.5%). Raised amylase in bile was observed in 8 .

Histopathological examination of all 36 patients (100%) revealed no evidence of malignancy.

Among 36 patients, surgical site infection was observed in 8 patients (22.2%), pulmonary complications in 6 patients (16.6%), cholangitis in 4 patients (11.1%), bile leak in 3 patients (8.3%), and anastomotic leak in 1 patient (2.8%). Re-exploration was required in 2 patients (5.55%), and 3 patients (8.33%) required readmission within one month.

Out of 36 patients, 20 patients (56%) had an uneventful recovery, while 9 patients (25%) developed Grade I complications, 5 patients (13%) had Grade II complications, and 2 patients (5.6%) developed severe complications corresponding to Grade IV A. At six months follow-up, incisional hernia was observed in 2 patients (5.6%), liver function derangement in 1 patient (2.8%), and anastomotic stricture detected on MRCP in 1 patient (2.8%).

Discussion

We found that the present study demonstrated a female preponderance (63.6%) with a median age of 38.5 years, which is consistent with the well-documented female dominance and adult presentation pattern of choledochal cysts. Similar observations have been reported by Todani et al. [8], who first classified choledochal cysts and noted that Type I and IV lesions are the most frequent, with a female predominance in adult cases. We also observed that right upper quadrant pain with mild jaundice was the most common presenting symptom (54.5%), followed by isolated pain and nonspecific gastrointestinal symptoms. These findings are comparable to the studies by Singh et al. [9] and Chandra et al. [10], where abdominal pain and jaundice were the predominant presenting features.

We observed that Type IV acholedochal cyst was the most common subtype (68.8%), followed by Type I (22.7%), which is in concordance with Wiseman et al. [11] and Komi et al. [12], who reported Type I and Type IV as the most frequent types in adult populations. The high incidence of Type IVa cysts further emphasizes the involvement of both intrahepatic and extrahepatic biliary systems. We also noted an anomalous pancreatobiliary junction (APBJ) in 13.8% of cases, which is a recognized etiological factor. This aligns with the findings of Babbitt [13], who highlighted APBJ as a key pathophysiological mechanism leading to pancreatic reflux and biliary epithelial injury.

We found that associated complications such as cholelithiasis (54.5%), recurrent pancreatitis (33.3%), and cholangitis (28%) were common in our cohort. Similar results were observed by O'Neill et al. [14], who reported high rates of gallstones and pancreatitis due to biliary stasis. We also observed secondary biliary cirrhosis with portal hypertension in 11.1% of patients, consistent with Tseng et al. [15], who described progressive hepatic fibrosis in untreated cases.

We found that all patients underwent choledochal cyst excision with Roux-en-Y hepaticojejunostomy, the standard surgical approach. This aligns with recommendations by Diao et al. [16], who emphasized complete cyst excision to prevent malignancy. In 5.55% of cases, additional pancreatic jejunostomy was required, consistent with Nagai et al., who advocated individualized management in patients with pancreatic involvement. We observed that postoperative recovery was satisfactory, with early return of bowel function and discharge. However,

complications such as surgical site infection, pulmonary complications, and bile leak were noted. These findings are comparable to Sharma et al. [17] and Yamaguchi et al., who reported similar postoperative morbidity profiles.

Conclusion

The present study highlights that choledochal cysts predominantly affect females in the adult age group, with right upper quadrant pain and jaundice being the most common clinical presentation. Type IVa was the most frequent morphological variant, and a significant proportion of patients had associated complications such as cholelithiasis, pancreatitis, and cholangitis, indicating the advanced nature of disease at presentation. Anomalous pancreatobiliary junction, although not very common, remains an important etiological factor contributing to disease progression. The standard surgical management with complete cyst excision and Roux-en-Y hepaticojejunostomy provided satisfactory outcomes in the majority of patients, with additional procedures required in select cases with pancreatic involvement. Postoperative recovery was generally favorable, with early return of gastrointestinal function and acceptable hospital stay. Although postoperative complications such as infection, bile leak, and pulmonary issues were observed, most were manageable, and severe complications were uncommon. Importantly, histopathological examination revealed no evidence of malignancy, reinforcing the role of timely surgical intervention in preventing malignant transformation. Follow-up findings indicate that while most patients recover well, a small proportion may develop late complications such as incisional hernia, liver dysfunction, or anastomotic stricture, underscoring the need for long-term surveillance. Overall, early diagnosis and complete surgical excision remain the cornerstone of management, offering good clinical outcomes and reducing the risk of long-term complications.

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