

# Presentation and Management of Choledochal Cysts in Children: The Experience of a Single Institute from a Developing Country

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## ABSTRACT

**Background:** Choledochal cysts are rare congenital biliary tract malformations. Their presentation varies based on the age and the type of cyst. Total cyst excision with biliary reconstruction is the treatment of choice. **Aim:** To present our experience with the presentation and management of these rare malformations with literature review. **Methods:** A retrospective analysis of a series of seven patients managed during the period from June 2015 to July 2020 in our institution is presented in this study. Recorded clinical data included sex, age at presentation, mode of presentation, diagnostic modality, cyst type and histopathology, preoperative course, operative procedure, and intraoperative and postoperative complications. **Results:** Of the patients enrolled in the study, 5 (71.4%) were female and 2 (28.6%) were male. Age at presentation ranged from 7 to 36 months. Abdominal pain was the most frequent manifestation and was recorded in 6 (85.7%) patients. The diagnosis was confirmed in all patients with MRCP. Cysts were type I in all (100%) patients. Open total cyst excision with Reux-en-Y hepaticojejunostomy was the standard procedure adopted in all patients. No significant intraoperative vascular or structural injury was recorded. Early postoperative complications were recorded in only 1 (14.3%) patient. All patients achieved good clinical outcome, and none showed significant findings on long-term follow-up. **Conclusions:** Choledochal cysts are rare congenital malformations with variable presentations. Total cyst excision with hepaticojejunostomy is a standard therapeutic option and is associated with an excellent outcome and decreased incidence of long-term risks.

**Keywords:** Children, choledochal cyst, hepaticojejunostomy, biliary ducts, malformations.

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## INTRODUCTION

A choledochal cyst is a rare congenital malformation that is characterized by single or multiple cystic dilatations of the intrahepatic and/or extrahepatic biliary ducts.<sup>1,2</sup> Halliday Douglas was the first to describe the pathology in 1852.<sup>3</sup> The overall incidence of choledochal cysts is estimated to be 1 in 100,000–150,000 live births in the Western population compared with 1 in 1,000 live births

in the Asian population, with a female predominance of 3–4:1.<sup>2,4</sup> Although reported with an increasing frequency among adults, 75% of choledochal cysts are still diagnosed during childhood.<sup>1,4</sup>

There is no general consensus regarding the etiology behind the development of choledochal cysts. The two most accepted theories suggest that the cystic dilatation

is either congenital secondary to an obstructing segment anywhere along the path of the biliary tree or acquired resulting from an anomalous pancreaticobiliary duct junction that could lead to the reflux of proteolytic pancreatic enzymes into the biliary tree with subsequent mural damage, weakness, and dilatation of the biliary ducts.<sup>1,3</sup>

According to Todani, choledochal cysts are classified into five types based on the anatomical findings and extent of biliary involvement. Type I cysts predominate and together with type IVa account for 90% of the cases.<sup>1,5</sup> Classically, choledochal cysts present with a triad of abdominal pain, jaundice, and abdominal mass; however, many reported that this triad is uncommon. Their presentation also differs based on the age of presentation and the type of the cyst. Abdominal mass and jaundice are common findings among infants, whereas abdominal pain is more frequently reported in older children.<sup>5,6</sup>

Ultrasonography is the imaging modality of choice in identifying biliary dilatation and has been well reported to be the initial imaging option in the diagnosis of these lesions.<sup>5,7</sup> Magnetic resonance cholangiopancreatography (MRCP) is highly sensitive in both detecting and classifying choledochal cysts with an overall detection rate of 96–100% and should be considered the first choice imaging technique for evaluating these malformations.<sup>5</sup> Total cyst excision along with biliary reconstruction is the treatment of choice. Drainage procedures without cyst excision are associated with an increased long-term risk of cholangitis, pancreatitis, and biliary tract cancer.<sup>6,8</sup>

Herein, we document our experience with the presentation and surgical management of choledochal cysts in children.

## MATERIALS AND METHODS

A retrospective analysis of a series of seven patients with choledochal cysts managed during the period from June 2015 to July 2020 in our institution is presented in this study. The study was approved by the Department of Pediatric Surgery Review Board in our institution. Recorded clinical data included sex, age at presentation, mode of presentation, cyst type and histopathology, preoperative course, operative procedure, and intraoperative and postoperative complications. The

diagnosis was confirmed in all patients by clinical history, physical examination, ultrasonography, and MRCP. Total cyst excision with biliary reconstruction through a Roux-en-Y Hepaticojejunostomy was the standard surgical procedure that was attempted in all patients.

## RESULTS

Of the patients enrolled in the study, 5 (71.4%) were female and 2 (28.6%) were male, with a female to male ratio of 2.5:1. Age at presentation ranged from 7 to 36 months with a mean of 20.7 months. Abdominal pain was the most frequent manifestation and was recorded in 6 (85.7%) patients, followed by jaundice in 3 (42.8%) patients. Abdominal mass was palpable in 2 (28.6%) patients. Vomiting was recorded in only 1 (14.2%) patient. The classic triad of (abdominal pain, jaundice, and palpable mass) was never reported in this study (Table 1). Abdominal ultrasonography was the initial imaging modality in all patients. The finding of cystic dilatation of the common bile duct was established in 4 (57.1%) patients. The diagnosis was confirmed in all patients with MRCP with an overall detection rate of 100% (Fig. 1). Cysts were type I in all (100%) patients (Table 1).

Preoperatively, 3 (42.8%) patients had elevated liver enzymes (AST and ALT), and 4 (57.1%) patients had elevated alkaline phosphatase. Direct hyperbilirubinemia was recorded in 3 (42.8%) patients. Vitamin K and plasma infusion were given over 3 days to correct prolonged prothrombin time in only 2 (28.6%) patients. None of the patients showed signs of preoperative cholangitis.

Total cyst excision with biliary reconstruction through a Roux-en-Y hepaticojejunostomy was performed in all (100%) patients (Fig. 2–6). No significant intraoperative vascular or structural injury was recorded. Mild biliary leakage was observed from the abdominal drain in only one patient and was managed conservatively with nasogastric decompression and intravenous antibiotics over a period of five days. Histopathological examination of all resected cysts revealed no malignant degeneration.

Duration of follow-up ranged from 12 to 38 months with a mean of 24 months, based on regular visits with clinical, biochemical, and ultrasonographic evaluation. None of the patients showed significant findings in terms of

cholangitis, abnormal liver function, and postoperative bowel obstruction.

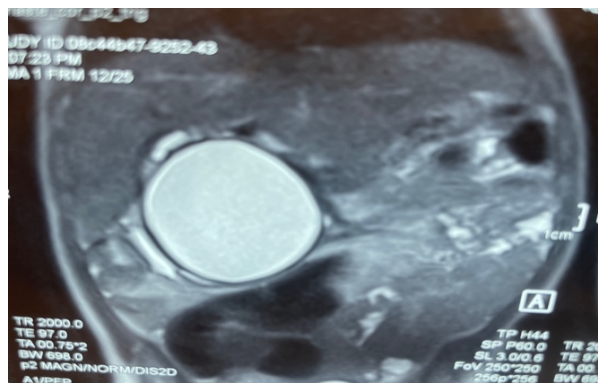


Figure 1: MRCP of a choledochal cyst type I in a 7-month-old female

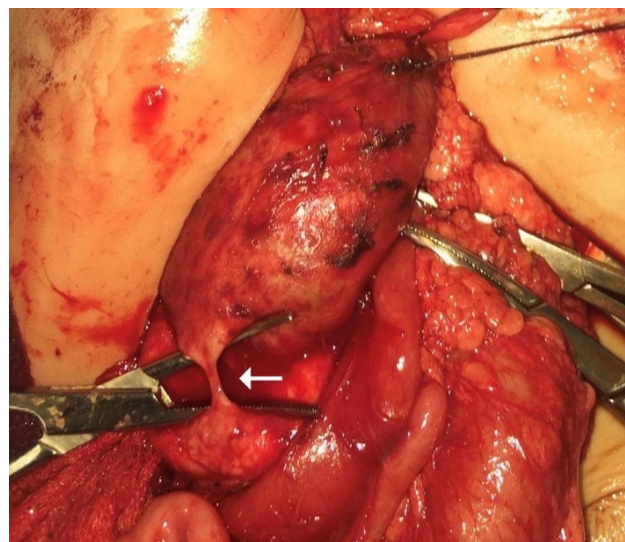


Figure 3: Dissection and division of the distal stenotic common bile duct (arrow).

Table 1: Demographic and clinical characteristics of patients.

Patient	Age (months)	Sex	Presentation	U/S	MRCP	Cyst Type
1	7	Female	Jaundice, Mass	Yes	Yes	I
2	12	Female	Jaundice, Pain	No	Yes	I
3	16	Female	Pain	No	Yes	I
4	19	Female	Pain	No	Yes	I
5	25	Male	Jaundice, Pain	Yes	Yes	I
6	30	Male	Pain, Mass	Yes	Yes	I
7	36	Female	Pain, Vomiting, Mass	Yes	Yes	I

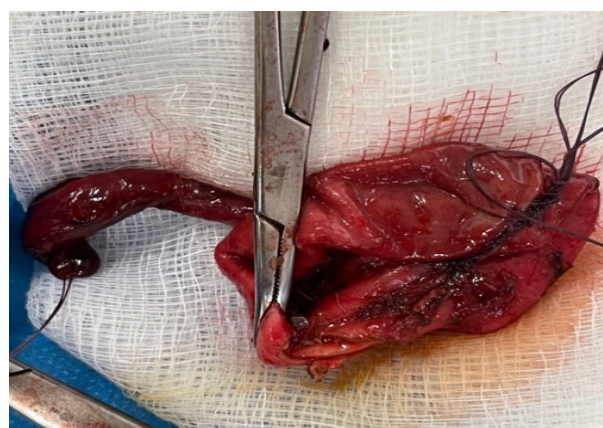


Figure 4: Total choledochal cyst excision

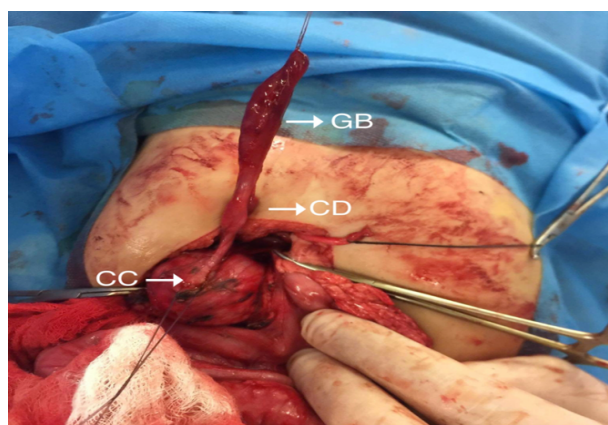


Figure 2: Operative demonstration: gall bladder (GB), cystic duct (CD), choledochal cyst (CC).

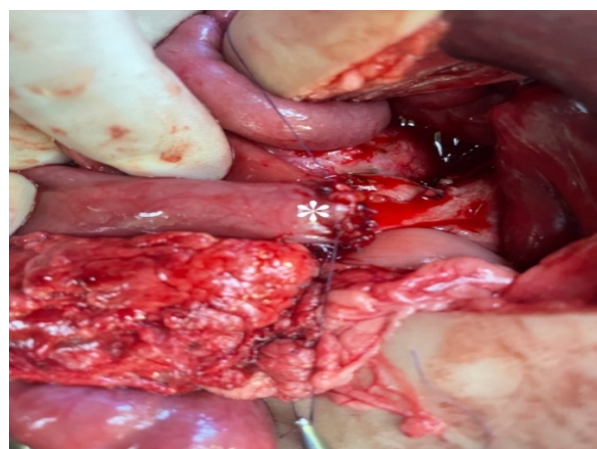


Figure 5: Hepaticojejunostomy (asterisk).





**Figure 6:** Retrocolic passage of the Roux-en-Y jejunal limb (arrow), Jejunojejunostomy (asterisk).

## DISCUSSION

The majority (71.4%) of patients enrolled in this study were female, with a female to male ratio of 2.5:1. This finding supports the observation that is well documented in the literature.<sup>2,4</sup>

There is no general consensus regarding the most frequent symptoms of choledochal cysts presenting in the pediatric age group. Some studies reported jaundice to be the leading symptom, while others reported abdominal pain as the most common manifestation.<sup>2</sup> In this study, abdominal pain was reported in 85.7% of the patients and was the most common presenting symptom. A similar observation was reported by Baez et al.<sup>4</sup> and Singham et al.<sup>9</sup> Although the classic clinical triad of abdominal pain, jaundice, and abdominal mass was reported in children with choledochal cysts, this association was not recorded in our study. A similar observation has been reported by others.<sup>2</sup> Clinical suspicion and the wide use of abdominal ultrasonography allowed early detection of choledochal cysts in infancy before they attained a large size to be abdominally palpable.

Abdominal ultrasonography was the initial imaging modality used in this study, and it was possible to establish the diagnosis in 57.1% of the patients. A similar observation was reported by others [2]. Other authors reported an ultrasound sensitivity of 94.7%.<sup>9</sup> In this study, abdominal ultrasonography suggested the diagnosis of simple liver cysts in two patients and pancreatic pseudocyst in one patient. We believe that being an operator-dependent technique, the younger age groups in our study and the rarity of the pathology

itself contributed to this observation. MRCP was used to ascertain the diagnosis in all patients in this study with an overall detection rate of 100%. This observation is consistent with what is reported by many others.<sup>2,5</sup>

According to Todani's classification, choledochal cysts type I predominate, and together with type IVa account for 90% of cases.<sup>1,5</sup> In this study, choledochal cysts type I were observed in 100% of the patients. Forny et al. and Singham et al. observed choledochal cysts type I in 93.4%, and 68% of their patients, respectively.<sup>2,9</sup> This finding may be attributed to the low sample size in our study in comparison with those in the mentioned studies.

Total cyst excision along with biliary reconstruction is the treatment of choice. Drainage procedures without cyst excision is associated with an increased long-term risk of cholangitis, pancreatitis, and biliary tract cancer.<sup>6,8</sup> Roux-en-Y hepaticojejunostomy and hepaticoduodenostomy are the two main options for biliary reconstruction following total cyst excision, and both can be carried out by open and laparoscopic approaches.<sup>6</sup> Recently, robotic-assisted techniques have been reported as the surgical option in the management of choledochal cysts in children.<sup>10</sup>

Open total cyst excision with Roux-en-Y hepaticojejunostomy was the standard procedure used in all patients in this study. Following cyst excision, the jejunum is divided distal to the ligament of Treitz, 20 cm in infants and 30 cm in older children, and the distal limb is brought up through a window in the transverse mesocolon to be anastomosed to the common hepatic duct. Bowel continuity is achieved by end-side jejunojejunostomy. Many surgeons still use hepaticojejunostomy following cyst excision. A continuous debate still exists whether hepaticoduodenostomy as a choice for biliary reconstruction is better or associated with fewer postoperative complications, and further studies are needed to achieve a solid conclusion.<sup>11</sup>

No significant intraoperative vascular or structural injury was recorded. Early postoperative complications were recorded in only one (14.3%) patient who had mild biliary leakage from the abdominal drain and was managed conservatively with nasogastric decompression and intravenous antibiotics over a period of five days. Forny et al. and Singham et al. reported early postoperative

complications in 13.3% and 31.3% of their case series, respectively.<sup>2,9</sup> All of the patients achieved good clinical outcome, and none showed significant findings in terms of cholangitis, abnormal liver function, and postoperative bowel obstruction on long-term follow-up.

Laparoscopic management of choledochal cysts in infants and children is considered a safe and feasible alternative.<sup>12</sup> In this study, all patients were surgically approached through an open technique. More experience is required through an advanced training to adopt a minimally invasive approach in the management of our patients safely in the future.

## CONCLUSIONS

A choledochal cyst is a rare congenital malformation with variable presentations based on age of presentation and type of the cyst. Total cyst excision with hepaticojejunostomy is a standard therapeutic option and is associated with an excellent outcome and decreased incidence of long-term complications.

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