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**ORIGINAL ARTICLE**

# Management Strategies for Choledochal Cysts in Infants and Children Our Experience at Department of Pediatric Surgery ,L.R.H, Peshawar

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## ABSTRACT

**Objective:** This study aims to understand more about choledochal cysts and how they are managed in children under 12 years old.

**Setting and Design:** A Descriptive Study at Department of Pediatric surgery L.R.H. Hospital Peshawar.

**Method and Material:** The most common kind of malformation of the biliary tree in Pakistan is choledochal cyst malformation (C.C.M.). Congenital choledochal anomalies range from asymptomatic to life-threatening cholangitis or pancreatitis. Surgeons now recommend total cyst ectomy and bilioentenric anastomosis. This five-year research examines. Clinical presentation, blood and laboratory testing, imaging, surgery, and predictive data were evaluated from case records. Twenty-three children with choledochal cysts were treated between January 2019 and December 2021, and their clinical presentation, diagnostic tests, and treatments were recorded. ERCP or a cholangiogram confirmed choledochal cysts after ultrasound diagnosis (P.O.C.). Before surgery, children with cholangitis received antibiotics and percutaneous transhepatic biliary drainage (excision of the cyst and jejunal loop interposition hepaticoduodenostomy)

**Results:** Twenty-three patients were analyzed; eight presented with chronic abdominal pain, five with a history of cholangitis, two with acute cholangitis, four with biliary peritonitis, and one with a history of pancreatitis. Following cyst excision, Roux-en-Y hepaticojejunostomy (RYHJ) was performed in 13 patients, and hepaticoduodenostomy (H.D.) was performed in 4 patients. One example of congenital choledochal malformation type II was treated with cyst excision and drainage because cholangitis and jaundice threatened the patient's life. The median age was 3, and the gender distribution was about even. Of the patients who sought care, 18 presented with jaundice, 15 with abdominal discomfort, 12 with fever, and 9 with a mass in their stomach. Only four people had all three symptoms of characteristic jaundice, pain, and lump. Overall, the results of both ERCP in 7 patients and P.O.C. in 14 cases were favorable. There were two distinct clinical presentations: I infantile form (less than a year), which included nine infants with jaundice in all, acholic stool in 7, lump abdomen in 4, but only one with classical triad; and (ii) childhood form (more than a year), which included twelve patients with abdominal pain, jaundice, and cholangitis. Twenty patients had a type I cyst, and three had a type Iva cyst. Only two kids said no to surgery, while the rest went through with it. Only three newborns were lost during surgery, but the other 18 did well following their follow-up visits (median 25 months). Six had extrahepatic biliary atresia, two had congenital hepatic fibrosis, and one had congenital biliary cirrhosis.

**Practical implication:** Our study will provide a new data to the health care providers about choledochal cysts and how they are managed in children under 12 years old

**Conclusions:** However, total cyst removal and Roux en Y hepaticojejunostomy (RYHJ) anastomosis is the definitive treatment of Congenital Choledochal Malformation. However, several other temporary therapies are also necessary at various phases of the illness. Their criticality—evolutionary stages may differentiate two types of choledochal cysts from infancy through adulthood. The prognosis is favorable following surgery, but early identification and referral are critical for reducing the risk of complications and mortality.

**Keywords:** Choledochal cyst, ERPC, H.D., RYHJ, and C.C.M. (Endoscopic retrograde cholangiopancreatography)

## INTRODUCTION

Choledochal cyst, localized aneurysmal dilatation of the extrahepatic and intrahepatic biliary system, is an uncommon etiology of obstructive jaundice that is often diagnosed in children[1]. Although choledochal cyst was initially described in print by Vater in 1723, it was not until 1852 that a patient with the condition was first described by name in a clinical setting. There have barely about 3,000 recorded instances between then and 1990. More than two-thirds of these cases are from India, and only two clinical series of choledochal cysts in children have been published so far[2]. One study is from over a decade ago3, and another compares children older than six years to adults, so we don't know what the whole clinical range of choledochal cysts in children looks like from either of these reports[3]. As a result, we looked at the symptoms, causes, and treatments for choledochal cysts in Pakistani kids under the age of 12[4]. It is customary to divide choledochal cysts into five distinct categories, delineated by their location5. Although several hypotheses and processes have been presented, the exact cause of choledochal cyst formation remains a mystery[5]. Approximately 70% of cases of acquired type have an aberrant pancreaticobiliary junction (APBJ) [6]. Choledochal cysts are often asymptomatic. However, they may sometimes manifest as biliary cirrhosis. In adolescents and teenagers, stomach discomfort most often occurs chronically or intermittently. Recurrent jaundice, cholangitis, biliary stones, pancreatitis, rupture of the common bile duct, and malignant transformation of the cyst8 are all possible complications of choledochal cysts[7]. There is concern that these kids will react terribly to a hepatotropic virus infection. Mildly increased liver enzymes, particularly gamma-glutamyl transferase (G.G.T.) and alkaline phosphatase, may serve as the first indicator of choledochal cysts. Diagnostic and categorization of choledochal cysts may be aided by endoscopic ultrasound, endoscopic retrograde cholangiopancreatography (ERCP), endoscopic ultrasound of the abdomen, computed tomography (C.T.), and magnetic resonance cholangiopancreatography (MRCP). In cases when prompt intervention is possible, the cyst may be surgically removed together with the biliary tree[8]. According to the literature, very limited data is available about the choledochal cysts and how they are managed in children under 12 years old. Therefore this study was carried out to understand more about choledochal cysts and how they are managed in children under 12 years old.

## METHODS & MATERIAL

We operated on 23 patients with choledochal cysts at MTI / LRH, Peshawar, between January 2019 and December 2021. Their medical history, including tests and treatments, was documented. All of them had an abdominal ultrasound. Diagnosis of choledochal cysts was verified by preoperative cholangiogram (P.O.C.) or

endoscopic retrograde cholangiopancreatography (ERCP) in the presence of a dilated common bile duct, with or without dilated intrahepatic ducts (ERCP). When portal hypertension was suspected clinically (splenomegaly) or sonographically, an esophagogastroduodenoscopy was performed, and if esophageal varices were seen, a second procedure was performed during follow-up. When medications failed to alleviate the symptoms of cholangitis in children, percutaneous transhepatic biliary drainage (PTBD) was performed11. Once the cholangitis was under control, all the kids were given the option to have surgery. The cysts were removed, and an interpositional jejunal loop hepaticoduodenostomy was performed during the surgical procedure. Portoenterostomy was performed on patients with choledochal cysts and extrahepatic biliary atresia (EHBA)12. A histologic analysis13 was performed on a wedge biopsy of liver tissue and a few other specimens. Todani et al. (s)14 categorized cysts into five groups: type I, a single extrahepatic cyst; type II, an extrahepatic supraduodenal diverticulum; type III, a choledococele; type Iva, both extra- and intrahepatic cysts; type IVb, numerous extrahepatic cysts; and type V, Caroli's illness (multiple intrahepatic cysts).

## RESULTS

The study period was from January 2019 to December 2021, and 23 patients with choledochal cysts were found in the pediatric surgery department at MTI / LRH, Peshawar. They had a median age of 3 (one month to 12 years). The ratio of men to women was 1.22 to 1. Symptom onset occurred anywhere from 3 days and nine years of age, and the duration of symptoms varied from 6 days to 7 years (median six months). In Table I, we can see a summary of their clinical characteristics. Cholangitis, stomach discomfort, and jaundice were the most common symptoms. Seventeen patients presented with the usual signs of abdominal discomfort, jaundice, and a mass in the stomach. Serum bilirubin, alkaline phosphatase, transaminases, and amylase were all abnormally high in 18 youngsters. Choledochal cysts would be detectable by ultrasound in any scenario. Seven instances (the youngest patient was 18 months old) were amenable to ERCP; in six of them, the results suggested Type I cysts, while one told a Type IVa cyst. Two of the six toddlers diagnosed with secondary biliary cirrhosis through preoperative wedge biopsy tragically passed away. One baby had regressed biopsy abnormalities on a repeat percutaneous biopsy eight months following surgery, whereas the other three were asymptomatic on the follow-up but still exhibited solid hepatomegaly. Four individuals were diagnosed with esophageal varices; in one child, the varices resolved shortly after surgery (perhaps owing to the release of pressure on the portal vein produced by the removal of the cyst), but in the other 3, they persisted during the subsequent follow-up (1 due to congenital hepatic fibrosis, two due to secondary biliary cirrhosis).

Table 1: Analyzing the Infantile and Juvenile Forms from a Clinical Perspective (Total Number of Patients 23)

| Patient's Characteristics | Overall | Infantile | Childhood |
|---|---|---|---|
| Median age (MO) | 36 | 5.5 | 63 |
| The median duration of symptoms (MO) | 6 | 2 | 20 |
| Jaundice | 18 | 9 | 9 |
| Pain abdomen/irritability | 15 | 3 | 12 |
| Fever | 12 | 4 | 8 |
| Lump abdomen | 9 | 4 | 5 |
| Classical triad | 4 | 1 | 3 |
| Cholangitis | 13 | 4 | 9 |
| Acholic stool | 7 | 6 | 1 |
| Secondary biliary cirrhosis | 6 | 4 | 2 |
| Stone in the cyst | 4 | 0 | 4 |
| Mean (SD) serum bilirubin (mg/dl) | 6.3 (5.9) | 10.8 (6.5) | 3.5 (3) |
| Liver histology | 23 | 14 | 9 |
| Type of cyst (Cystic | 19 | 11 | 8 |
| Type of cyst (Fusiform) | 16 | 0 | 16 |
| Sex (male) | 7 | 3 | 4 |
| Sex (female) | 16 | 5 | 11 |
| Age | | 0 to 8 months | 2 to 12 years |
| Biliary Amylase | | + | ++++++ |

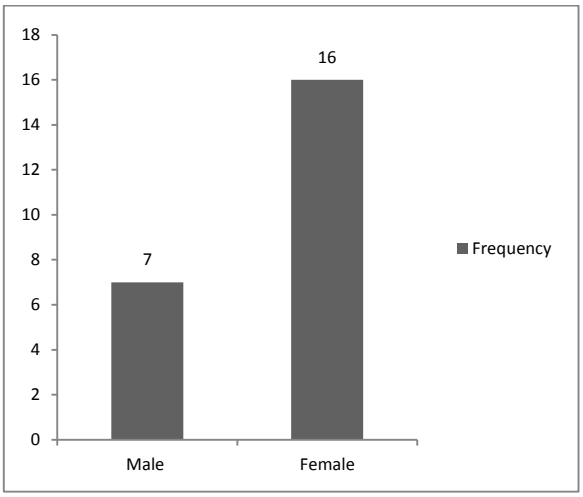


Figure 1: Gender wise distribution of the participants

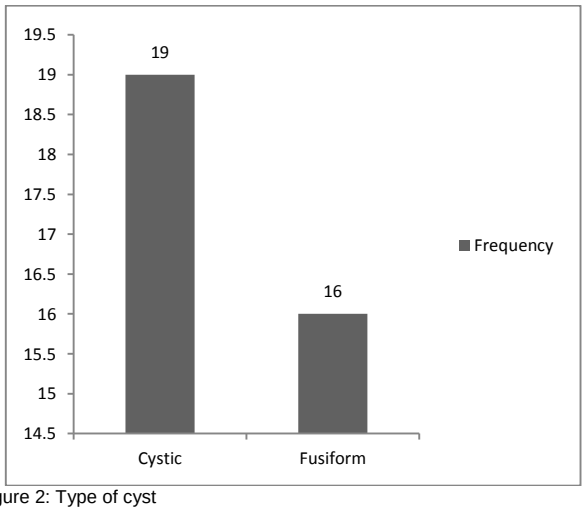


Figure 2: Type of cyst

## DISCUSSION

We share our understanding and experience in managing choledochal cysts in children of various ages. Most pediatric patients report intermittent nonspecific stomach pain 15. However, 11 babies and newborns did so with jaundice, acholic stools, and an abdominal tumor[9]. Most choledochal cysts in children had a fusiform morphology, whereas those in newborns were often cystic[10]. Twenty of the twenty-three juvenile patients had elevated biliary amylase, and this was connected with an aberrant pancreaticobiliary junction. Neonatal bile duct obstruction, liver fibrosis, and cirrhosis were prevalent[11].

To sum up, choledochal cysts are not the same as biliary atresia (B.A.) and should not be treated like that. Minimal morbidity may be expected with early excision of choledochal cysts and biliary repair in neonates, babies, and children. Choledochal cysts are a very uncommon congenital dilatation of the biliary ducts18 (Incidence; 1:10,001–1:150,000 live births) [12]. Biliary lithiasis, cancer, chronic liver injury, portal hypertension, and cirrhosis19 may develop if delayed treatment. Choledochal cysts may appear at any age; however, in 80-90% of cases, the diagnosis is made before 20[13]. The extensive use of ultrasonography in evaluating

nonspecific abdominal pain in children and obstetric presentation, pathology, surgical strategy, and prognosis at various pediatric ages21 have all contributed to a decrease in the age of diagnosis[14]. The clinical data (sex, age at presentation, and clinical manifestations), laboratory investigations (serum bilirubin-direct and indirect, SGOT, gamma G.T., alkaline phosphatase; (biliary amylase levels), and liver histology, HIDA SCAN operative procedure, and outcome were reviewed for 23 children admitted for the Management of choledochal cyst over three years. The patient's characteristics are summarized in table 1[15]. Females were up the majority (78%), and presentation ages spanned from 0 to 16 years23. Newborn cysts had a cystic morphology, whereas most cysts in children aged six and above were fusiform. There were very few cases with the "classical trio" of jaundice, discomfort, and abdominal tumor[16]. There was an increase in biliary amylase. In group II, 24 patients had abnormalities in the pancreaticpancreaticobiliary junction on the preoperative cholangiogram.

In contrast, P.O.C. showed abstract thought at the low end of CBD in newborns. Regarding aberrant liver histology, eight individuals in group I had portal fibrosis, and 3 had biliary cirrhosis[17]. Upon diagnosis, all patients had immediate choledochal cyst removal and enteric biliary repair. Five patients experienced complications between 6 months and 15 years, including one with a wound infection, two with biliary leakage, one with pancreatitis, and two with cholangitis[18]. Definitive surgery is based on removing the whole enlarged extrahepatic bile duct without causing damage to other essential tissues such as the portal vein, hepatic artery, pancreas, and its duct, or duodenum[19]. Before the ductal hole can be sealed, the protein plugs/sludge must be removed from the common channel[20].

## CONCLUSION

The complicated Choledochal Cysts Management (C.C.M.) management necessitates careful preparation and a multimodality approach. Depending on the particulars of each diagnosis, the right course of treatment must be determined. Percutaneous or endoscopic biliary drainage operations might supplement the surgical treatment. Positive results are seen in patients whose complex situations have been handled correctly. In light of the potential for severe, even fatal, consequences during Choledochal Cysts Management, prompt surgical intervention is warranted (C.C.M.). Due to the significant risk of postoperative cholangitis, intraoperative flushing of the hepatic duct should be reserved for specific instances.

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