



# Unusual presentation of a choledochal cyst

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## Introduction

Choledochal cysts are a rare phenomenon, with the incidence in western countries estimated to be between 1:100,000 to 1:150,000 births [1]. However, the incidence may be higher in other countries, with some reports suggesting up to 1:1000 [2]. They are more common in females, with a ratio of 3:1 or 4:1 [3,4]. Due to their congenital predominance, most choledochal cysts have been diagnosed in the pediatric population [5]. Their pathogenesis remains unclear, although recent studies suggest that an abnormal pancreaticobiliary junction (ABPJ) may play a role [6]. Choledochal cysts often present with many other associated anomalies, including: Biliary atresia, bowel atresia (duodenal, jejunal, etc.), imperforate anus, multiseptate gallbladder, and others. Specifically, choledochal cysts appear to be the result of an unequal proliferation of embryologic biliary epithelial cells before duct cannulation is complete [7,8]. Gen-

erally, patients present with the classic triad of abdominal pain, jaundice, and a palpable mass [9]. However, this presentation is far more common in children than adults. Furthermore, neonates usually present with obstructive jaundice and abdominal masses [10]. In this report, we describe a 4 month old female with a choledochal cyst presenting with jaundice and neonatal hyperbilirubinemia.

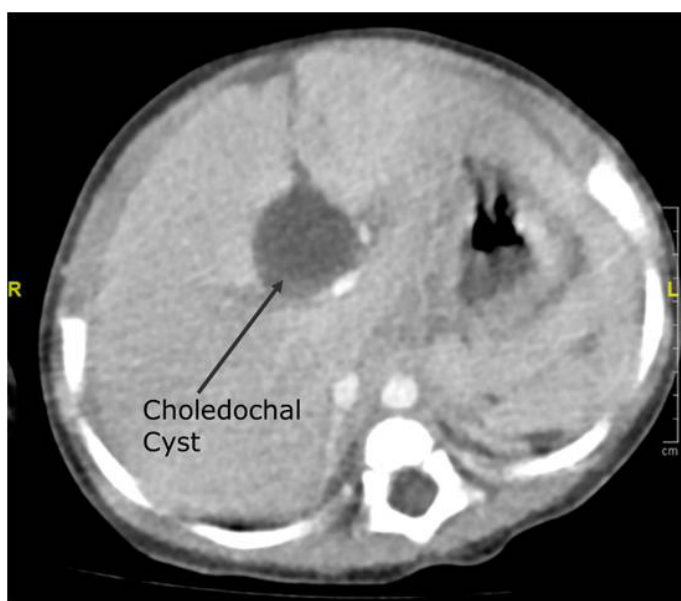
## Case Presentation

A 4 month old female was referred to the pediatric surgery clinic for icteric sclera and severe jaundice. Birth history was significant for uncomplicated C-section delivery, 1 week stay in NICU. Complications in the NICU included transient Tachypnea of The Newborn (TTN), mild Respiratory Distress Syndrome (RDS), neonatal hyperbilirubinemia, mild right atrial enlargement, pul-



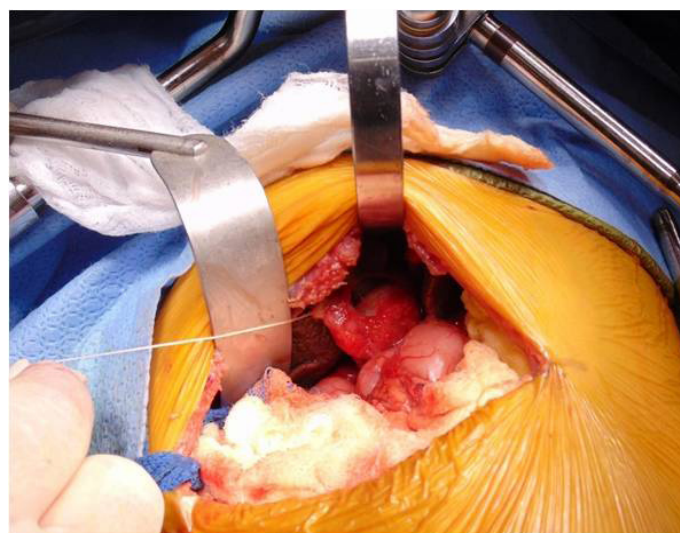
monary regurgitation, dilated coronary sinus, right aortic arch, and a reducible umbilical hernia. Family history was significant for neonatal jaundice in two older siblings that resolved naturally. The patient had normal feeding habits, a normal number of stools, and a normal number of wet diapers. All other histories were noncontributory. Bilirubin levels were trended in the NICU, with total bilirubin ranging from 10.6-15.5 and direct bilirubin ranging from 1.2-1.5. Based on these values and the possibility of physiologic neonatal jaundice, phototherapy was initiated immediately after birth, and decreased total bilirubin to 11.1 and direct bilirubin to 1.0.

Upon presenting to the pediatric surgery clinic, a repeat set of bilirubin labs was drawn, showing 9.4 total, 5.2 direct, and 4.2 indirect. At this time, AST (386.1), ALT (234.1), and Alkaline Phosphatase (643.9) were also ordered, with all values above the normal ranges. Coagulation tests were ordered as a follow up, showing an INR of 1.82, PT of 19.3, and PTT of 42.2, all above the normal ranges. Imaging was also initiated, with hepatic ultrasound showed a 2.7x1.8cm cystic mass in the common bile duct that appeared to communicate with the gallbladder. All other anatomy was within normal limits. A follow up abdominal CT scan demonstrated a cyst in the porta hepatis (Figure 1). Based on these findings, the patient was scheduled for an exploratory laparotomy with cystectomy, intraoperative cholangiogram, and Roux-En-Y choledochojejunostomy



**Figure 1:** CT of Abdomen showing cyst in porta hepatis

Prior to surgery, the patient was transfused 120mL FFP and one unit Vitamin K to assist with pre-operative care. At surgery, the choledochal cyst did not communicate with the duodenum, instead ending as a blind pouch (Figure 2). Additionally, the liver was extremely cirrhotic (Figure 3). Furthermore, the choledochal cyst communicated with the intrahepatic bile duct and there was anatomical variance of the left lobe of the liver. Based on these findings, the diagnosis was modified to extrahepatic biliary atresia, an unusual variant of a blind ending choledochal cyst.



**Figure 2:** Choledochal cyst ending as blind pouch

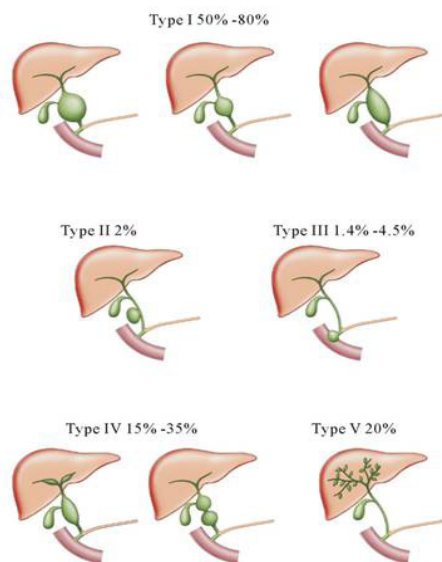


**Figure 3:** Cirrhotic Liver (see arrow)

Post-operative complications included acute respiratory failure for which patient was placed on a ventilator. Additionally, laboratory testing on POD#1 showed persistent high liver function tests (AST 763, ALT 395, ALP 513) and a total bilirubin of 11.6. Thus, the patient was transferred to Children's Medical Center in Dallas for further treatment with hepatology.

## Discussion

The current classification scheme for biliary cysts is based on the 2003 guidelines to include extrahepatic, intrahepatic, and ABPJ cysts, an addition to the Todani classification of 1977 (Figure 4) [3,11-13]. Type I cysts, which make up 50-85% of total cases, encompass cystic or fusiform dilations of the common bile duct. Type II cysts (2% of cases) are true diverticula of the extrahepatic bile ducts and communicate with the bile duct through a narrow stalk. These may arise from any portion of the extra hepatic bile duct. Type III cysts (1-5% of cases) are cystic dilations limited to the intraduodenal portion of the distal common bile duct (commonly known as choledochal cysts). Type IV cysts (15-35% of cases) describe the presence of multiple cysts. Type V cysts (20% of cases) include one or more cystic dilations of the intrahepatic ducts, without any extrahepatic duct disease. A common variant is Caroli Disease. Lastly, type VI cysts (exceedingly rare) describe isolated cystic dilations of the cystic duct, to date only described in case reports.



**Figure 4:** Todani Classification of biliary cysts

When formulating a differential diagnosis for jaundice (and thus neonatal hyperbilirubinemia) one can divide the disorder into those of increased production and those of decreased clearance. Increased production disorders include isoimmune mediated hemolysis, inherited erythrocyte membrane defects (hereditary spherocytosis, elliptocytosis), erythrocyte enzymatic defects (G6PD deficiency, pyruvate kinase deficiency), and sepsis. On the other hand, many newborns present with disorders of decreased bilirubin clearance. These include direct bilirubinemias such as physiologic jaundice of the newborn (most common cause, treated with phototherapy and resolves in 1-2 weeks), biliary tract obstructions, and genetic defects such as Dubin-Johnson syndrome and Rotor syndrome. Indirect bilirubinemias also exist, including Crigler-Najjar (Type I and II) syndrome and Gilbert syndrome.

Currently, multiple treatment modalities exist for management of neonatal hyperbilirubinemia secondary to a choledochal cyst. Most commonly, symptomatic choledochal cysts are excised surgically, either through endoscopic resection or via sphincterotomy [14]. This procedure is usually accompanied by a Roux-En-Y hepaticojejunostomy in order to provide a route for biliary drainage from the liver [15,16]. While this is usually a successful procedure, the most common post-operative complication is stenosis of the biliary-enteric anastomosis leading to cholangitis, jaundice, or cirrhosis. In cases of concurrent biliary atresia, the Kasai procedure is often performed, which involves a hepatopertoenterostomy designed to restore bile flow from the liver to the small intestine [17]. If successful, patients will begin to see resolution of their jaundice within several weeks following the procedure. Unfortunately, for the majority of patients that do not experience resolution, liver transplant evaluation must be considered within three months of the procedure.

#### Learning points/take home messages

1. It is crucial to identify patients with congenital choledochal duct cysts and/or concurrent developmental abnormalities in order to best choose method of treatment
2. Multiple treatment modalities exist, but all should be evaluated with care to best suit the patient and their individual circumstances
3. In many cases, liver transplant may be the only true

method of long term success in biliary atresia

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