

# Another Type of Choledochal Cyst Beyond the Todani Classification

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**The classification of choledochal cyst has changed from 3 types described by Alonso-Lej to 5 types described by Todani, but these classifications do not include dilatation of the cystic duct. The sixth type of choledochal cyst has been described by some authors for the cystic dilatation of the cystic duct. It is so rare that accurate diagnosis is difficult before operation. We present a 15-month-old girl with a type VI choledochal cyst that was misidentified preoperatively as a type I choledochal cyst. Besides the gall bladder and the cystic duct cyst, we also excised the distal part of the common bile duct and performed reconstruction with Roux-en-Y hepaticojejunostomy. From a review of the literature, we have determined that there is a more direct route to adequate diagnosis and management of the sixth type of choledochal cyst.**

*Key words:* Biliary duct – Choledochal cyst – Cystic duct

A choledochal cyst is defined as a congenital dilatation of the bile duct, and its incidence is reported to be approximately 1 in 100,000 to 150,000.<sup>1</sup> These cysts more commonly develop in women. Choledochal cysts can be found at any age, but almost two thirds of patients exhibit symptoms in the first decade of life. The classification has changed from the 3 types described by Alonso-Lej to 5 types described by Todani, but these classifications do not include dilatation of the cystic duct. Some authors<sup>2–4</sup> refer to this entity as the sixth type of choledochal

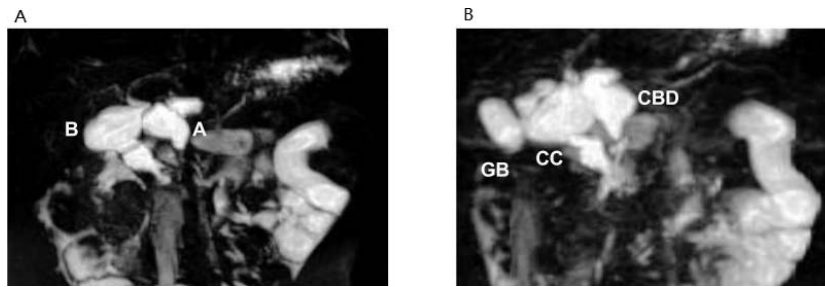
cyst. There have been fewer than 16 cases reported in the English literature, mostly in sporadic case reports.<sup>2,3</sup> We present a case of a type VI choledochal cyst and review the literature to determine the basis of diagnosis and optimal management.

## Case Presentation

A 15-month-old girl presented with poor appetite and frequent postprandial vomiting; she had been

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**Fig. 1** (A) In this preoperative MRCP, B was thought to be the gall bladder, and A was thought to be a fusiform cystic dilatation of the common bile duct. A type I choledochal cyst was suspected. (B) In another view of the preoperative MRCP, the relationship between the cystic duct cyst (CC), and the gall bladder (GB), and common bile duct (CBD) was clearer.

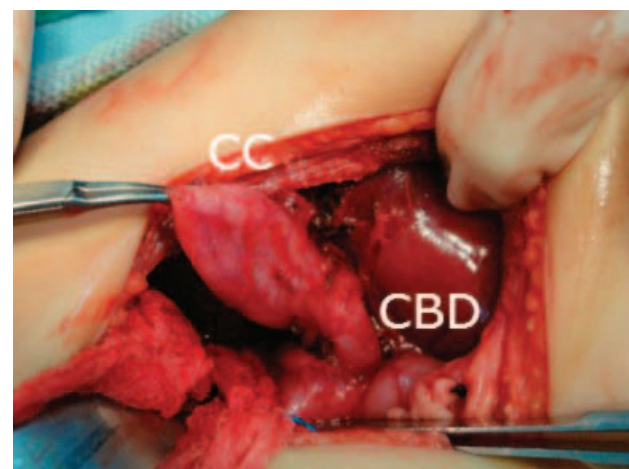
previously healthy and had met major developmental milestones. Initially, conservative therapy was administered for a presumptive diagnosis of acute gastroenteritis. After 5 days of this approach, an intermittent high fever was noted, yet there was no definite source of infection. She was then transferred to our Pediatric Emergency Department for further workup. The physical exam revealed epigastric and right upper abdominal tenderness without muscular guarding or rebound tenderness. Laboratory studies revealed increased liver function test parameters [aspartate aminotransferase (AST) 977 U/L and alanine aminotransferase (ALT) 712 U/L] and increased serum amylase (226 U/L) and serum lipase (2700 U/L) levels. Mild leukocytosis (white blood cell count of 11,700/ $\mu$ L) without a left shift was also observed. Conventional abdominal ultrasound showed a dilated extrahepatic bile duct without stones or mass lesions. A coronal magnetic resonance cholangiopancreatogram (MRCP) demonstrated a fusiform dilatation of the common bile duct up to 1.6 cm in diameter involving most or all of the extrahepatic duct (Fig. 1A). There was also abnormal insertion of the common bile duct into the pancreatic duct with a common channel of approximately 1.1 cm in length. A type I choledochal cyst was diagnosed, and surgical management was planned.

During the operation, a fusiform cystic lesion was encountered first, and it communicated with the common bile duct with a wide opening (Figs. 2 and 3). The gall bladder diameter was within normal limits, and it drained into the cyst through a narrow duct. The middle part of the common bile duct revealed mild dilatation. Aspiration from the cystic lesion before excision was performed to determine the amylase level (970 U/L). We performed a cholecystectomy as well as excision of the cyst and a segment of the common bile duct as distally as possible. Reconstruction of the biliary continuity was performed with a Roux-en-Y hepaticojejunostomy. The patient was discharged without any specific

morbidities on postoperative day 7. Pathology reports showed that the cystic duct was dilated with thickened fibrous walls and lined with single-layered to pseudostratified, benign-appearing biliary epithelium. There were no dysplastic changes or malignant features observed in the gall bladder, cystic duct, or common bile duct specimens. At the 2 follow-ups at postoperative months 3 and 6, the patient was healthy. Future annual follow-ups are planned.

## Discussion

Cystic malformation of the cystic duct was first suggested as another type of biliary cyst by Serena Serradel *et al.*<sup>4</sup> It has been reported as isolated cystic duct dilatation or cystic duct dilatation associated with other findings, including a fusiform dilatation of the common bile duct.<sup>5,6</sup> The most common symptom in these patients is pain located in the epigastric area or right upper abdomen; obstructive jaundice or acute cholangitis develops in some of these cases.<sup>2</sup> In the present case, abdominal pain was the only related



**Fig. 2** As shown in this intraoperative photograph, the gall bladder was excised, and a fusiform cyst of the cystic duct (CC) was noted with a wide opening into the common bile duct (CBD).



**Fig. 3** A diagrammatic sketch with the specimen was used to illustrate the dilatation of the cystic duct and its relationship to the gall bladder and common bile duct.

finding. Furthermore, pancreatitis and abnormal liver functions were the key laboratory abnormalities in this case. An accurate preoperative diagnosis of type VI choledochal cyst seems to be difficult because it is a rare entity and exhibits ambiguous anatomic features. Most cases are misidentified as type I or II choledochal cysts before the operation. Yoon first made a proper diagnosis preoperatively for 3 patients with the use of MRCP.<sup>7</sup> Praveen Maheshwari<sup>5</sup> correctly preoperatively identified 10 cases using abdominal Doppler ultrasonography combined with MRCP or multidetector computed tomography (MDCT). Once any nonvascular cystic structure is found close to the porta hepatis, its relationship with the gall bladder, cystic duct, and common bile duct should be thoroughly determined.<sup>5</sup> There are 2 types of type VI choledochal cyst based on the cases reported by Praveen Maheshwari<sup>5</sup>; these include saccular and fusiform dilatations of the cystic duct. It is possible to differentiate the saccular type of cystic duct dilatation from type II choledochal cysts only when there is a narrow or normal cystic duct between the cyst and the common bile duct.<sup>5</sup> The condition might be misdiagnosed as a type I choledochal cyst when fusiform dilatation of the common bile duct is also present.<sup>6</sup> We reexamined the MRCP results in this case and discussed the findings with our radiologist. In addition to the wide opening to the common bile duct, the fusiform dilatation of the cystic duct was extremely close to the dilated common bile duct as in Mirrizi syndrome. In this situation, it was easy to mistake the cyst as a type I choledochal cyst. After we reexamined the image using the method suggested by Praveen

Maheshwari, we clearly determined the connections among the gall bladder, cystic duct cyst, and common bile duct (Fig. 1B).

Several mechanisms are thought to be related to the formation of type VI choledochal cyst. Based on the unique anatomic abnormalities of type VI choledochal cysts, an abnormal pancreaticobiliary duct junction (APBDJ) is the most likely and accepted mechanism.<sup>6</sup> There are 3 components of the APBDJ hypothesis: (1) acute angulation of the common hepatic duct and cystic duct junction; (2) reflux of the mixed pancreatic and bile juice; and (3) stasis. All result in cystic duct ectasia, and then dilatation eventually develops. A high amylase level in the cyst supports the APBDJ hypothesis and was observed in our case. A retrospective study revealed a different relationship between the biliary amylase level and clinical features of choledochal cysts; there might be another pathophysiologic basis other than the APBDJ hypothesis.<sup>8</sup> Other mechanisms have been proposed as the cause of this condition; one theory states that a dilatation originates from the weakest part of the duct resulting from the lowest vascularity of the biliary tree, focal aganglionosis such as Hirschsprung disease, or microlithiasis.<sup>6</sup> None of the above theories provides a comprehensive explanation of the pathogenesis of type VI choledochal cysts, so more cases and further analyses are required.

Traditionally, choledochal cysts can be adequately managed with cyst excision with or without hepaticojejunostomy according to the cyst type. The appropriate management of type VI choledochal cysts is determined based on the cystic duct opening into the common bile duct.<sup>2,5,6</sup> Cholecystectomy with simple cyst excision is feasible for isolated cystic duct dilatations with narrow or normal cystic ducts between the cyst and the common bile duct. Cystic duct cysts with wide openings to the common bile duct are usually accompanied by an abnormal common bile duct. Therefore, excision of the cyst en bloc with the gall bladder and distal common bile duct is appropriate for cases involving a wide cystic duct cyst opening into the common bile duct. Malignant neoplasms of the biliary tract develop in 2.5% to 28% of patients with choledochal cysts, and the risk of malignancy increases with age.<sup>9</sup> Furthermore, patients with choledochal cysts are diagnosed with cholangiocarcinoma most commonly at 32 years of age, which is 20 years earlier than in the general population.<sup>9</sup> Regardless of the selected procedure, the pathology reports should carefully determine whether biliary

intra-epithelial neoplastic changes are present. A second operation with extensive excision should be considered in cases involving a simple excision of a cystic duct cyst and a subsequent discovery of a premalignant lesion.<sup>6</sup> Although there is no evidence of the incidence of malignancy in type VI choledochal cysts following surgery, several reports have revealed an approximately 0% to 6% incidence of malignancy following surgery for the other types of choledochal cysts.<sup>9</sup> In addition to early follow-up after the operation, regular postoperative follow-up is important.

In conclusion, it is difficult to identify type VI choledochal cysts preoperatively. However, there are some signs related to their anatomic features based on diagnostic modalities, which should be considered if there is a high index of suspicion by surgeons and radiologists. The cystic duct orifice and the diameter of the common bile duct are important for determining adequate management during the operation. To adequately address any cancer risk, a detailed pathologic examination of specimens should be implemented, especially in adult patients. Although type VI choledochal cysts are still uncommon, more cases might be reported in the future as clinicians become increasingly familiar with their characteristics.

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