

## Choledochocele Presenting as Recurrent Pancreatitis

Erica Kaye, Sara Mixer, Ketan Sheth M.D., and Kitt Shaffer M.D., Ph.D.

We report the case of a 31-year-old male with recurrent episodes of acute pancreatitis, subsequently discovered to have a rare type III choledochal cyst, also termed a choledochocele. This case demonstrates the utility of multiple imaging techniques to diagnose the correct etiology of the patient's pancreatitis, as well as to appropriately plan surgical intervention. For many years, endoscopic retrograde cholangiopancreatography has been the gold-standard for diagnosis of type III choledochal cysts; this procedure, however, carries a significant degree of morbidity and may perhaps be circumvented with the advent of advanced imaging techniques that allow for visualization of the intraduodenal portion of the biliary tract. In this case, CT and MR imaging demonstrated a spherical, cyst-like structure extending from the pancreatic duct into the second part of the duodenum, suggestive of a choledochocele. Presence of the choledochocele and its exact anatomy were confirmed with ERCP. This imaging, in combination with the appropriate clinical constellation of symptoms, enabled correct identification of the etiology of the patient's unexplained recurrent episodes of pancreatitis, allowing for appropriate and curative surgical intervention.

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Abbreviations: CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography; MRI, magnetic resonance imaging

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### Case Report

A 31-year-old man presented to the Emergency Department with complaint of one day of severe, sharp, mid-epigastric pain without radiation to other regions of the abdomen. The pain awakened him from sleep, and it continued to worsen over the course of the day, with episodes of severe cramping that lasted for several minutes and spontaneously resolved. He denied pain associated with eating, although he reported feeling "gassy" following consumption of food. He had been eating and drinking fluids regularly since the onset of pain. He denied nausea, vomiting, diarrhea, constipation, fever, chills, or headache. He denied any similar symptoms in

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the past, and was not aware of any prior history of gallstones. His past medical history was significant only for various dental procedures, and was otherwise noncontributory. He was taking ibuprofen and hydrocodone/acetaminophen as needed for pain, and had no known drug allergies. His family history is significant for a father who died of gastric cancer, type unknown. There is no other history of gallstones or any other stomach, intestinal, pancreatic, or liver problems in his family. He did not use tobacco, rarely drank alcohol, and denied use of recreational drugs.

On physical exam, the patient was found to be afebrile, with normal cardiac rate and rhythm, blood pressure, and respiratory rate. His sclerae were anicteric. His abdomen was tender to palpation at the midline epigastric region into the right upper quadrant, with questionable guarding in the right upper quadrant. Neither rebound tenderness nor distention was noted. The liver was slightly enlarged. There was no evidence of periumbilical or flank discoloration, nor any other regions of ecchymosis. Neurological and extremity exams were normal. Laboratory investigation revealed white blood cell count and electrolytes within normal limits, as well as elevated lipase at 306 (normal 20-120 U/L), elevated amylase at 295 (normal 20-140 U/L), slightly elevated AST 74 (normal 0-40 U/L) and ALT 54 (normal 0-40 U/L), and normal alkaline phosphatase and total bilirubin. Initial abdominal radiograph and right upper quadrant ultrasound were ordered, and both were read as normal. In retrospect, however, the outline

of an enlarged gallbladder is visible in the right upper quadrant on the radiograph (Fig. 1). The ultrasound is remarkable for an enlarged gallbladder with thickened wall and pericholecystic fluid (Fig. 2A), as well as dila-



Figure 1. 31-year-old man with recurrent pancreatitis and choledochocoele. Initial abdominal radiograph shows enlarged gallbladder (arrow).

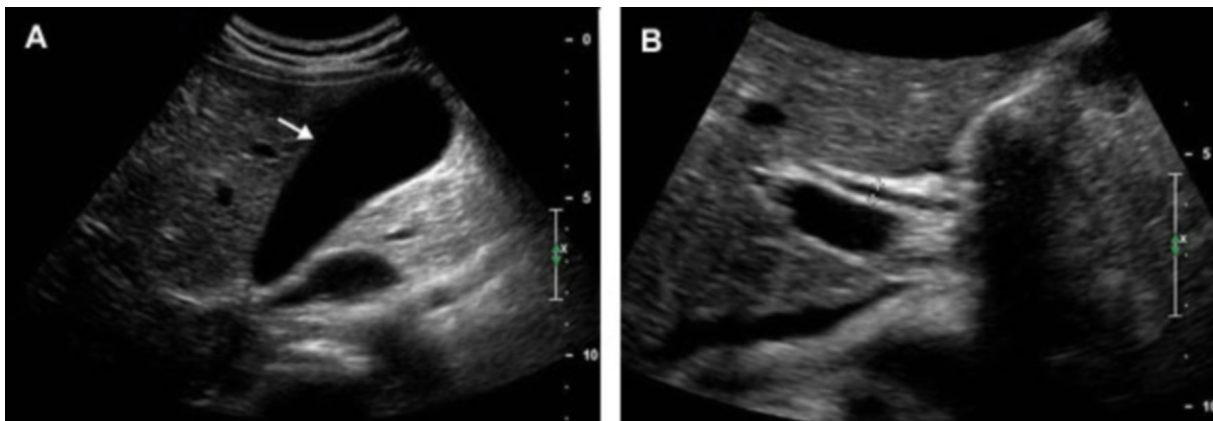


Figure 2A-B. 31-year-old man with recurrent pancreatitis and choledochocoele. Initial abdominal sonogram demonstrates (A) an enlarged gallbladder with thickened wall and increased pericholecystic fluid (arrow), and (B) dilated common bile duct (cross-hatches).

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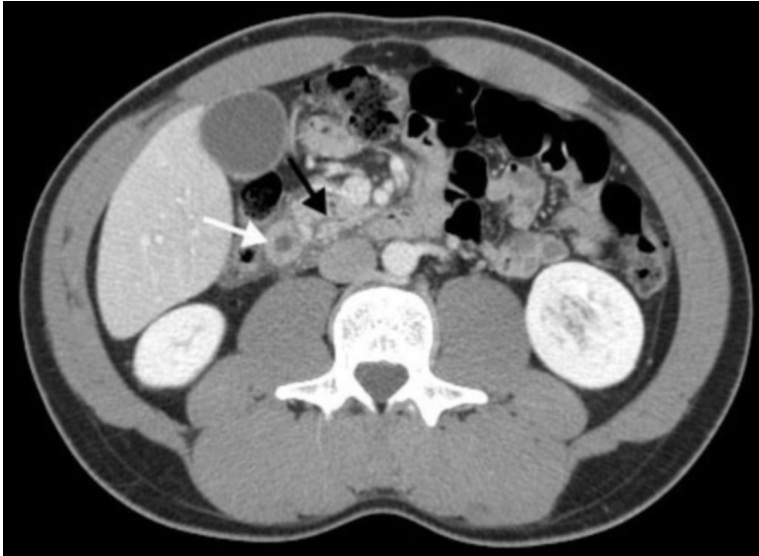


Figure 3. 31-year-old man with recurrent pancreatitis and choledochocele. Axial CT demonstrates 7 mm cystic structure within duodenal lumen (white arrow) and pancreatic duct (black arrow) communicating with cyst.

tion of the common bile duct (Fig. 2B). The patient's constellation of abdominal symptoms in the context of elevated pancreatic enzymes was deemed consistent with pancreatitis, with precise etiology unknown at the time of admission.

After admission, the patient was aggressively rehydrated with intravenous fluids and given patient-controlled analgesia with morphine for pain control. Further investigation of the etiology of the pancreatitis included CT of the abdomen and pelvis, which demonstrated a 7 mm intraduodenal cystic structure extending from the pancreatic duct into the second portion of the duodenum (Figs. 3-4), most consistent with a type III choledochal cyst. There was a small amount of free fluid in the right upper quadrant, with mild periportal edema, however the gallbladder was unremarkable. There was no evidence of bowel obstruction, free air, appendicitis, or diverticulitis. The pancreatic enhancement pattern was unremarkable, as were the spleen, kidneys, and surrounding vascular structures.

In an effort to confirm the diagnosis, MR cholangiopancreatography (MRCP) of the abdomen was subsequently performed using axial T1 and SPGR, as well as coronal and axial single shot T2-weighted sequences

with thin and thick slab MIP images of the biliary tree (Fig. 5). The scan was limited by respiratory motion artifact and low spatial resolution. The scan showed no dilatation of the biliary tree and distention of the gallbladder, with no evidence of gallstones to explain the pancreatitis. There was a trace of pericholecystic fluid, unchanged from the recent CT imaging. The pancreatic duct was not dilated. However, the choledochal cyst suggested by the CT was not evaluated well by this study, and its existence could neither be confirmed nor excluded.

As a result of these findings, the patient was diagnosed with acute pancreatitis secondary to intraduodenal choledochocele. The patient was managed expectantly with continued intravenous fluids and ketorolac for pain control, and his symptoms re-

solved after 2 days. He was discharged following a total of 5 days as an inpatient, and given instructions to avoid use of ibuprofen and to decrease dietary intake of fatty or fried foods. He was also instructed to follow up with gastroenterology in six to eight weeks, with recommendation to undergo endoscopic retrograde cholangiopancreatography (ERCP) to further define the anatomy of the cyst.

Per patient report, he adhered strictly to a low-fat diet but continued to experience intermittent episodes of acute, severe, abdominal pain. Four days following his discharge, the patient returned to the Emergency Department with complaint of recurrence of abdominal pain. Clinical presentation and laboratory values at this time (amylase and lipase values greater than 300) were consistent with a recurrence of his pancreatitis. Over the next week, he reported to his primary care practitioner twice with complaint of severe epigastric pain that was exacerbated by eating and drinking and not alleviated by high doses of hydrocodone and later morphine. After this second visit, immediate referral was made for the patient to undergo ERCP, which took place five days later. ERCP visualized an ampulla that resembled an egg projecting into the duodenal lumen, and the ampullary

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opening of the cyst was too small for a standard catheter to be used for the injection of contrast into the biliary tree. Once the ducts were visualized, cystic dilatation at the beginning of the pancreatic and bile ducts was observed, consistent with a type III choledochal cyst (Fig. 6). There was also a mild cystic dilatation of the left hepatic duct within the liver. The patient developed worsening of his abdominal pain, nausea, and vomiting after the ERCP, consistent with exacerbation of his pancreatitis. At this point, he was referred to surgery for evaluation.

Following the recommendation of the surgeon, the patient underwent open abdominal surgery to resect the choledochocele, with concomitant prophylactic cholecystectomy. The cyst was accessed via the duodenum. Once the cyst was visualized, it was apparent that the cyst itself had a very small ampullary orifice, such that a probe could not be passed through it (Fig. 7A). The cyst wall was opened, and there were 2 visible openings,

one for the pancreatic duct and one for the common bile duct, draining directly into the cyst (Fig. 7B). The cyst wall was amputated and the septum between the two ducts partially divided, allowing passage of a probe into both orifices and biliary and pancreatic secretions to emanate freely. Following a four-day postoperative recovery period, the patient was discharged from the hospital. He has recovered well and is being followed by gastroenterology.

### Discussion

Choledochal cysts encompass a range of anatomical anomalies characterized by cystic dilatation in various regions of the biliary tract. The Alonso-Lej classification of choledochal cysts is based on the specific location of the cystic dilatation, refined further by Todani et al. [1] to include 5 major types of choledochal cysts:

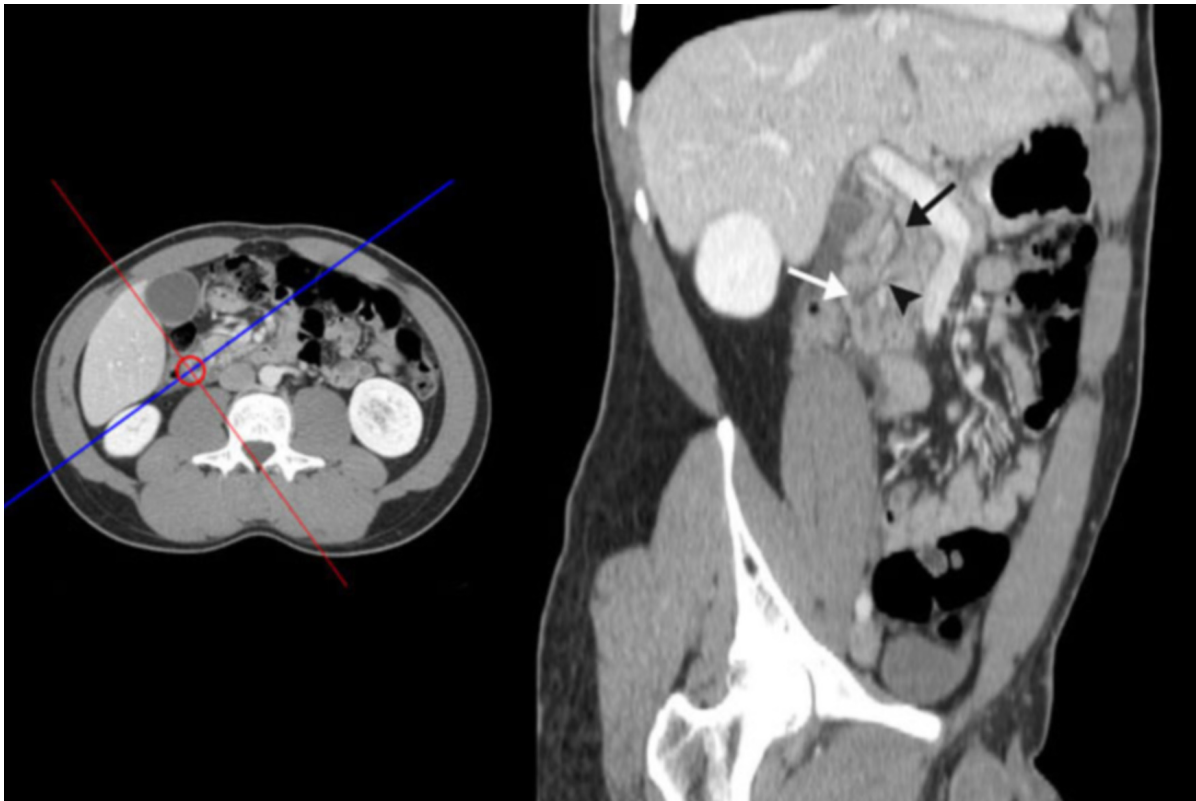


Figure 4. 31-year-old man with recurrent pancreatitis and choledochocele. Reformatted oblique sagittal CT shows common bile duct (black arrow) and pancreatic duct (arrow head) in direct communication with cyst (white arrow).

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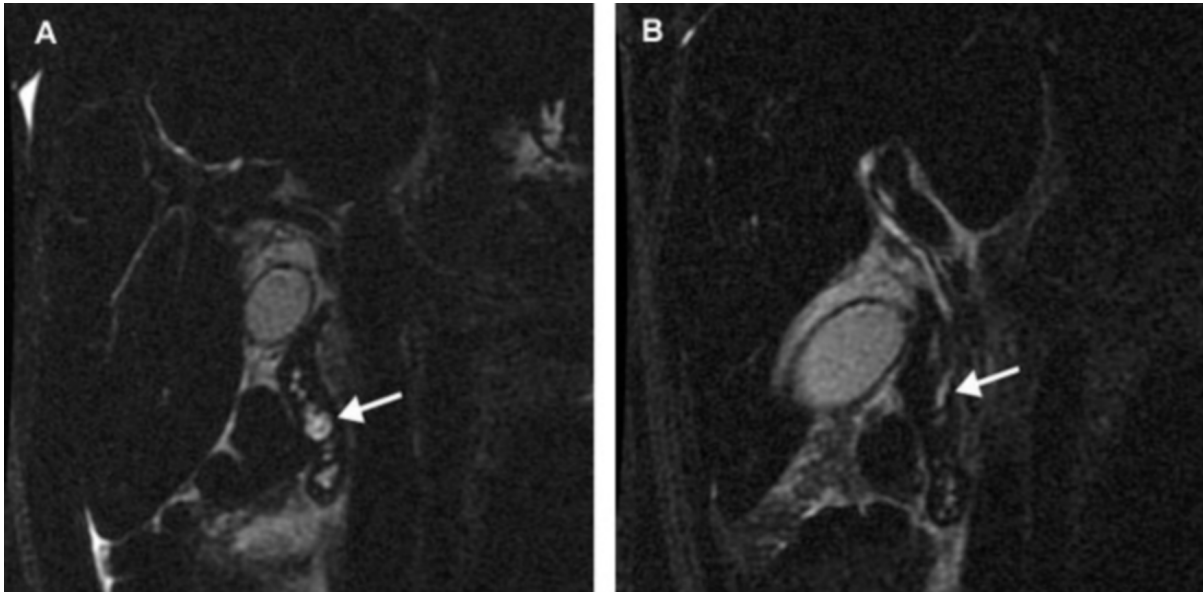


Figure 5A-B. 31-year-old man with recurrent pancreatitis and choledochocoele. Reformatted oblique MRCP shows (A) intraluminal duodenal cyst and (B) pancreatic duct (arrows).

- Type I cysts are the most common type, representing 80-90% of all lesions, and are characterized by dilatation of the entire common hepatic and common bile ducts, or of segments of each; subtypes a, b, and c represent differences in cystic appearance, such as saccular or fusiform.
- Type II cysts are characterized by isolated protrusions or diverticula projecting from the common bile duct wall; they may be further classified into sessile or pedunculated subtypes.
- Type III cysts, also known as choledochocoeles, are characterized by existence of the cystic component within the intraduodenal portion of the common bile duct. Choledochocoeles are further classified as type IIIa (intraluminal with common opening for the common bile duct and pancreatic duct), type IIIb (intraluminal with separate openings for the common bile duct and pancreatic duct), and type IIIc (completely intramural).
- Type IV cysts are characterized by multiple dilatations of the intrahepatic and extrahepatic biliary tree, with type IVA involving both intra- and extrahepatic dilations and type IVB involving only extrahepatic dilations.
- Type V cysts, otherwise known as Caroli Disease, are characterized by intrahepatic biliary dilation, without extrahepatic involvement [1,2].

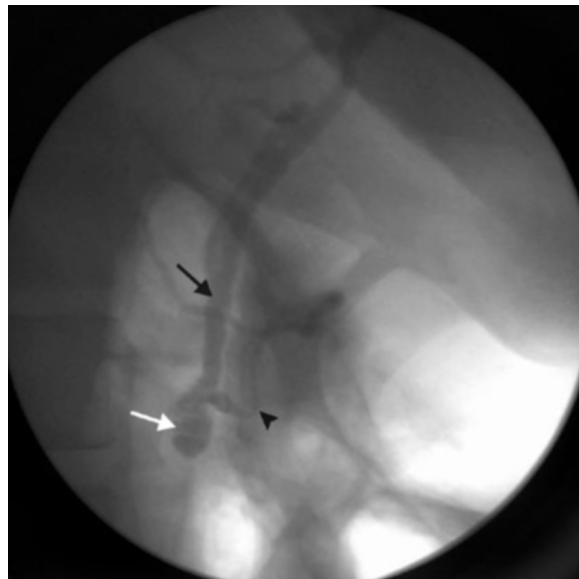


Figure 6. 31-year-old man with recurrent pancreatitis and choledochocoele. ERCP demonstrates intraluminal choledochal cyst (white arrow) direct communication with dilated common bile duct (black arrow) and pancreatic duct (arrowhead).

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Choledochal cysts occur relatively rarely in the United States and other Western countries, with incidence ranging from 0.00005% to 0.001%. The incidence is higher in Asia, with 33% of all reported cases occurring in Japan, where Miyano and Yamataka have reported a prevalence of as high as 0.1% [3]. The choledochocele (type III) is one of the least common types, occurring in only 1.4 to 4.5% of all cases of choledochal cysts [4].

The majority of patients with choledochal cysts present during infancy or early childhood, with 80% of cases diagnosed prior to age 10 years [5]. Over the past several decades, however, this disease has been increasingly recognized in adults, although rarely with the classic childhood symptomatic triad of jaundice, abdominal pain, and an abdominal mass [6]. The majority of adults present with complaints of vague epigastric or right upper quadrant pain [7]. Critical morbidities associated with choledochal cysts include pancreatitis, cholangitis, and cholangiocarcinoma, with the rate of malignant degeneration of a choledochal cyst ranging from 9-28%. In patients who have choledochal cysts at 10 years of age or younger, the risk of developing cholangiocarcinoma is 1%, whereas the risk increases to 15% for patients older than 20 years of age [8].

The large majority of choledochal cysts are congenital, with a very rare number of acquired cases discussed in the literature. The pathogenesis of choledochal cysts is thought to be multifactorial, with various components interacting to produce congenital weakness of the biliary duct wall. In addition, genetic defects in the critical

processes of epithelialization and recanalization of the developing bile ducts occurring during organogenesis may contribute to the formation of congenital cysts. Choledochal cysts often occur in the context of contiguous anomalous junction of the common bile duct with the pancreatic duct, otherwise known as anomalous pancreatobiliary junction. In one study, Miyano and Yamataka demonstrate anomalous pancreatobiliary junction occurring in greater than 90% of patients with choledochal cysts [3]. Anomalous pancreatobiliary junction is thought to play an etiologic role in the development of choledochal cysts, particularly those associated with duodenal obstruction [9]. In an effort to evaluate pathogenesis of choledochal cysts, an experimental model of anomalous pancreatobiliary junction was surgically engineered in laboratory canines; various degrees of common bile duct dilatation was shown to develop in 100% of canines within 7-10 days, with no further dilatation occurring in subsequent days [10]. Histological analysis revealed evidence of chronic pancreatitis in several of the subjects, indicating that anomalous pancreatobiliary junction may be an important etiologic factor not only in formation of choledochal cysts, but also in the pathogenesis of chronic pancreatitis, a known consequence of choledochal cyst disease.

Although ERCP has long been regarded as the gold standard for diagnosing choledochal cysts and evaluating anomalous pancreatobiliary junction, over the past ten years multiple studies have shown MRCP to be just as sensitive as, if not more sensitive than, conventional

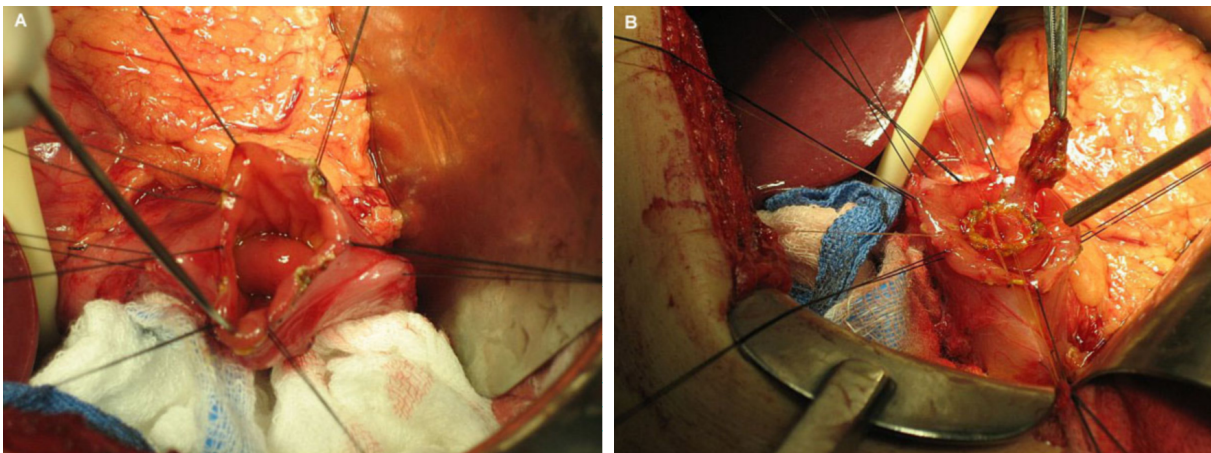


Figure 7A-B. 31-year-old man with recurrent pancreatitis and choledochocele. Intraoperative photos show cyst within the duodenum (A) and, after cyst resection, the orifices of the common bile and pancreatic ducts (B).

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cholangiogram [11, 12]. Since MRCP is a noninvasive study, it further avoids some of the significant morbidity associated with ERCP, including moderate to severe pancreatitis, seen in this patient's case as well as almost 2% of patients undergoing ERCP [13]. MRCP, however, is not without its limitations, including the potential for serious compromise by movement artifact, as demonstrated in this case. Resolution also presents a limitation in the case of small cysts, as seen in this case and in some pediatric cases of choledochal cyst. The scan in this case was performed with a slice thickness of 1 cm, just larger than the diameter of the cyst itself; therefore, even under ideal conditions MRCP likely would not have provided the necessary anatomical detail of the cyst and pancreaticobiliary junction.

Another advantage of ERCP over MRCP is that ERCP offers the potential of therapeutic intervention. Cyst wall excision with or without sphincterotomy can be performed during ERCP in the case of most small, uncomplicated choledochoceles. Both procedures have been found to be curative of the obstruction caused by the cyst with minimal complications, and circumvent major surgical intervention with its associated risks [14]. In this case, the gastroenterologist was concerned that the cyst was too large and potentially too complex for intervention during ERCP. However, understanding the precise anatomy of the cyst and pancreaticobiliary junction was essential for planning surgical intervention to correct the choledochocele. Since this information had not been attained through less invasive imaging, ERCP was still necessary in order to definitively image the anomaly. ERCP showed conclusively that the patient's pancreatic and biliary ducts emptied directly into the cyst itself, as opposed to being directly adjacent to the cyst, and that there was minimal cystic involvement of either duct. With this information, surgical intervention could be limited to excision of the intraluminal portion of the cyst wall, with no need to reconstruct the ducts themselves.

For this patient, a variety of imaging modalities in synergy with the clinical presentation ultimately elucidated a rare etiology of recurrent pancreatitis, allowing for appropriate and curative surgical intervention.

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