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Single Balloon Enteroscopy-Assisted ERCP for Treatment of Cholangitis in a Patient with a Kasai Portoenterostomy

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Abstract

Primary therapy for biliary atresia is a surgical hepatoportoenterostomy (Kasai procedure), which has been shown to reduce mortality, but is frequently complicated by ascending cholangitis and the development of biliary cirrhosis. Previously reported therapy for recurrent cholangitis caused by biliary obstruction has included surgical revision and percutaneous biliary drainage, but ERCP has not been previously described. Here we report a patient with recurrent cholangitis following a Kasai procedure and an anastomotic stricture successfully treated with single balloon enteroscopy-assisted ERCP. This novel technique could be considered in patients with this common complication of the Kasai procedure, and may impact long-term outcomes in this patient population.

Keywords

ERCP; Kasai; Portoenterostomy; Single Balloon Enteroscopy

INTRODUCTION

The Kasai procedure (hepatoportoenterostomy) is the primary treatment for infants with biliary atresia, an obliterative cholangiopathy of the extrahepatic bile ducts that can result in biliary fibrosis and cirrhosis. The surgery is performed by creation of an anastomosis between a Rouxen-Y loop of bowel and the liver hilum to restore bile flow to the small bowel. Multiple series have demonstrated substantial improvements in mortality with this procedure, but long-term complications remain a significant problem, with the majority of patients developing cholangitis and liver failure, and often requiring liver transplantation. Previously reported treatment of recurrent biliary obstruction and cholangitis has been limited to antibiotics, corticosteroids, percutaneous transhepatic cholangiography (PTC), and surgical revision. These therapies are limited by suboptimal efficacy of the medical treatments and the morbidity associated with the percutaneous and surgical approaches. Endoscopic retrograde cholangiopancreatography (ERCP) is generally the first-line therapy for bile duct strictures but in the past was not available for patients with altered Roux-en-Y

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anatomy. However, recent advances in deep enteroscopy have allowed endoscopic access to the bile ducts of patients with Roux-en-Y anatomy following gastric bypass and liver transplant. Single balloon enteroscopy, which is commonly used to evaluate and treat small bowel pathology, is performed with a flexible fiberoptic endoscope fitted with an overtube and an inflatable balloon that allows anchoring and "pleating" of the small bowel to attain deep intubation (Figure 1). The enteroscope itself is not designed specifically for ERCP, but devices designed for biliary cannulation and intervention can be passed through the enteroscope. ERCP using the single balloon enteroscope has not been described for patients with a prior Kasai procedure. Here we describe the use of single balloon enteroscopy-assisted ERCP to treat a biliary stricture in a patient with a prior Kasai who developed recurrent cholangitis.

CASE REPORT

A 19 year-old woman was admitted to the hospital for her fourth episode of cholangitis in four years. She was diagnosed in utero with an intrahepatic choledochal cyst and underwent cyst resection with hepaticocholecystostomy at 16 weeks of age. Intraoperative cholangiography showed patency of the intrahepatic bile ducts and emptying of the gallbladder into the duodenum via a patent common bile duct. At 30 weeks, she presented with jaundice and acholic stool, with hepatobiliary scintigraphy demonstrating uptake within the liver and a lack of radiotracer within the main hepatic ducts, gallbladder, and duodenum. At laparotomy, the porta hepatis was found to have dense adhesions and fibrosis, and examination of the gallbladder showed a patent, intact anastomosis without bile flow proximally. Cholecystectomy was performed, followed by open liver biopsy, showing marked intracanalicular cholestasis with bile thrombi, bile ductular proliferation, scattered inflammation of the hepatic parenchyma, and periportal fibrosis. Based on this information, the treating physicians diagnosed the patient with acquired biliary atresia.

At age 34 weeks, she was readmitted with vomiting and a 5 cm biloma near the porta hepatis. Percutaneous drainage was performed with continued drainage of bile over the following days, prompting repeat exploration with planned portoenterostomy. Dissection of the porta revealed scant bile drainage from small ducts of the left lobe and none from the right lobe. A Roux-en-Y portoenterostomy confined to the left hepatic lobe was performed, with subsequent clinical improvement and normalization of the serum bilirubin.

She continued to do well until age 15, when she began to develop recurrent cholangitis necessitating repeated hospitalizations for intravenous antibiotics. She was also found to have portal hypertension, with gastroesophageal varices and splenomegaly. A liver biopsy showed portal expansion, bile ductular proliferation, cholestasis, and stage 3 biliary fibrosis. Despite medical therapy with ursodiol and chronic suppressive oral antibiotics, she continued to have recurrent episodes of cholangitis. Doppler ultrasonography documented normal flow through the hepatic artery on multiple occasions. During one hospitalization at age 19, MRCP was performed showing marked biliary dilatation with hypertrophy of the left hepatic lobe (Figure 2). Due to the Roux-en-Y anatomy, a single balloon enteroscopy-assisted ERCP was performed. A single balloon enteroscope was used to achieve endoscopic access to the portoenterostomy, where a biliary orifice was not readily apparent (Figure 3A). Close observation revealed an area of mucosa where bile slowly pooled, and this area was probed with a guidewire followed by a balloon catheter (Figure 3B). Contrast injection via the catheter confirmed positioning within a bile duct (Figure 4A). The orifice was balloon dilated to 10 mm (Figure 3C). Following dilation, an adjacent orifice was identified which was cannulated and confirmed to be another left hepatic duct branch (Figure 4B). This orifice was then dilated to 8 mm (Figure 3D). Moderate dilatation was seen within both of the ducts, with adequate drainage following dilation on delayed fluoroscopic imaging. The

total serum bilirubin decreased from 6.1 to 3.4 mg/dl following the procedure, and there have been no further episodes of cholangitis after 8 months of follow-up.

DISCUSSION

Ascending cholangitis occurs commonly in patients following hepatoportoenterostomy. The mechanism for this complication is not entirely clear, but likely contributors include intestinal bacterial stasis and incomplete biliary drainage. Afferent limb intestinal obstruction at the mesocolic window has also been reported to cause cholangitis. In addition to its immediate morbidity, episodes of cholangitis have been associated with worse outcomes and progression of portal hypertension, although this association has not been consistently reported. To address this frequent problem, intestinal decontamination through the use of prophylactic antibiotics and mechanical relief of partial biliary obstruction through the use of PTC and surgical revision have been utilized. The use of adjuvant corticosteroids to inhibit the inflammatory response has also been used, but a clinical benefit has not been demonstrated in controlled studies.

The use of a single balloon enteroscope to facilitate ERCP in patients with Roux-en-Y anatomy has been previously reported, but these series did not include patients with a prior Kasai procedure. To our knowledge, this is the first report of endoscopic treatment of a biliary stricture in a patient with a prior Kasai procedure. Admittedly, the initial diagnosis of biliary atresia in this patient is highly questionable, given the patient's unusual history and multiple surgeries prior to the Kasai. Clearly she did not have congenital biliary atresia. However, it is possible that she suffered an iatrogenic bile duct injury during the choledochal cyst excision. Such an injury could result in an inflammatory obliteration of the distal duct, resulting in “acquired biliary atresia.” One previous description of acquired biliary atresia included an infant with an iatrogenic bile duct injury during duodenal surgery, resulting in severe fibrosis and adhesions in the porta hepatis, similar to our case. Despite the uncertainty of the initial diagnosis in this case, her post-Kasai course of recurrent cholangitis with associated biliary fibrosis and portal hypertension is commonly seen in the typical patient with a prior Kasai. Additionally, if dilated bile ducts are seen on imaging, we would expect that patients with a Kasai performed for classic congenital biliary atresia would benefit similarly from endoscopic dilation of an anastomotic biliary stricture. Indeed, successful dilation of strictures using this technique has been reported for patients with a hepaticojejunostomy. Finally, for those cases caused by an obstruction of the afferent limb, single balloon enteroscopy would be able to diagnose the obstruction and potentially treat it with balloon dilation.

Relief of biliary obstruction in patients following the Kasai procedure may improve patient outcomes by preventing recurrent cholangitis, and could attenuate progression of liver disease and portal hypertension. Although surgical revision and PTC have been shown to be effective for this purpose, endoscopic therapy has the advantage of being less invasive and without the need for external drainage. This technique is effective as well, with a 70% success rate in one large series of patients with Roux-en-Y anatomy. Most of the failed attempts were due to inability to reach the biliary anastomosis as a result of failure to identify the afferent limb, bowel angulation, and inability to enter the afferent limb. Thus, the length of the afferent limb may not have a large impact on the success rate, although an unusually long afferent limb may preclude a successful procedure. In addition to its effectiveness, single balloon enteroscopy is safe, with a low complication rate among procedures performed for gastrointestinal luminal disease, and no complications reported when performed for ERCP. Replication of this technique, particularly in patients with classic biliary atresia, and assessment of long-term outcomes are needed to establish ERCP as a standard treatment modality for this patient population.

Acknowledgments

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Abbreviations

PTC	Percutaneous transhepatic cholangiography
ERCP	Endoscopic Retrograde Cholangiopancreatography
MRCP	Magnetic Resonance Cholangiopancreatography

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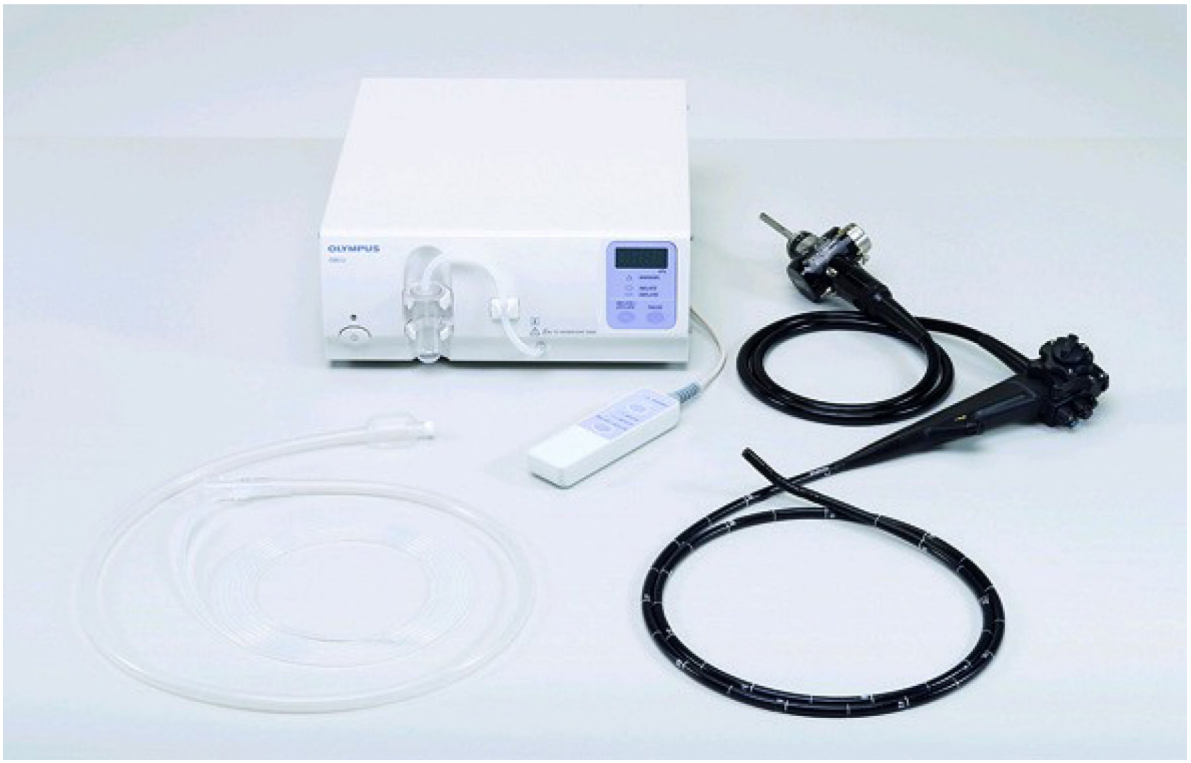


Figure 1.
Single balloon enteroscopy system. Endoscope pictured on the right and balloon-fitted overtube on the left. The overtube balloon is inflated and retracted to pleat the small bowel, and the endoscope is advanced. This technique is repeated multiple times to achieve deep intubation of the small bowel. (Image courtesy of Olympus America, Inc.)

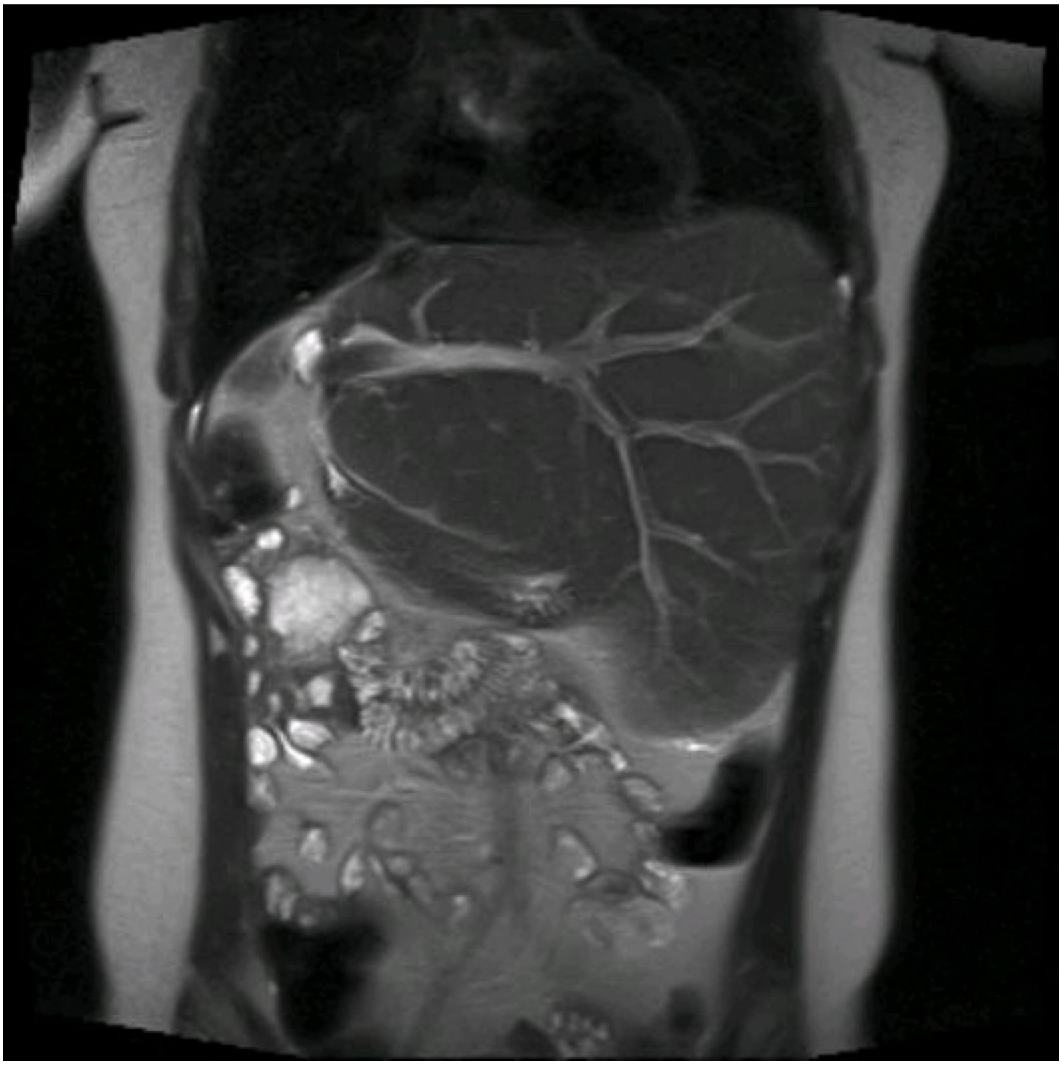
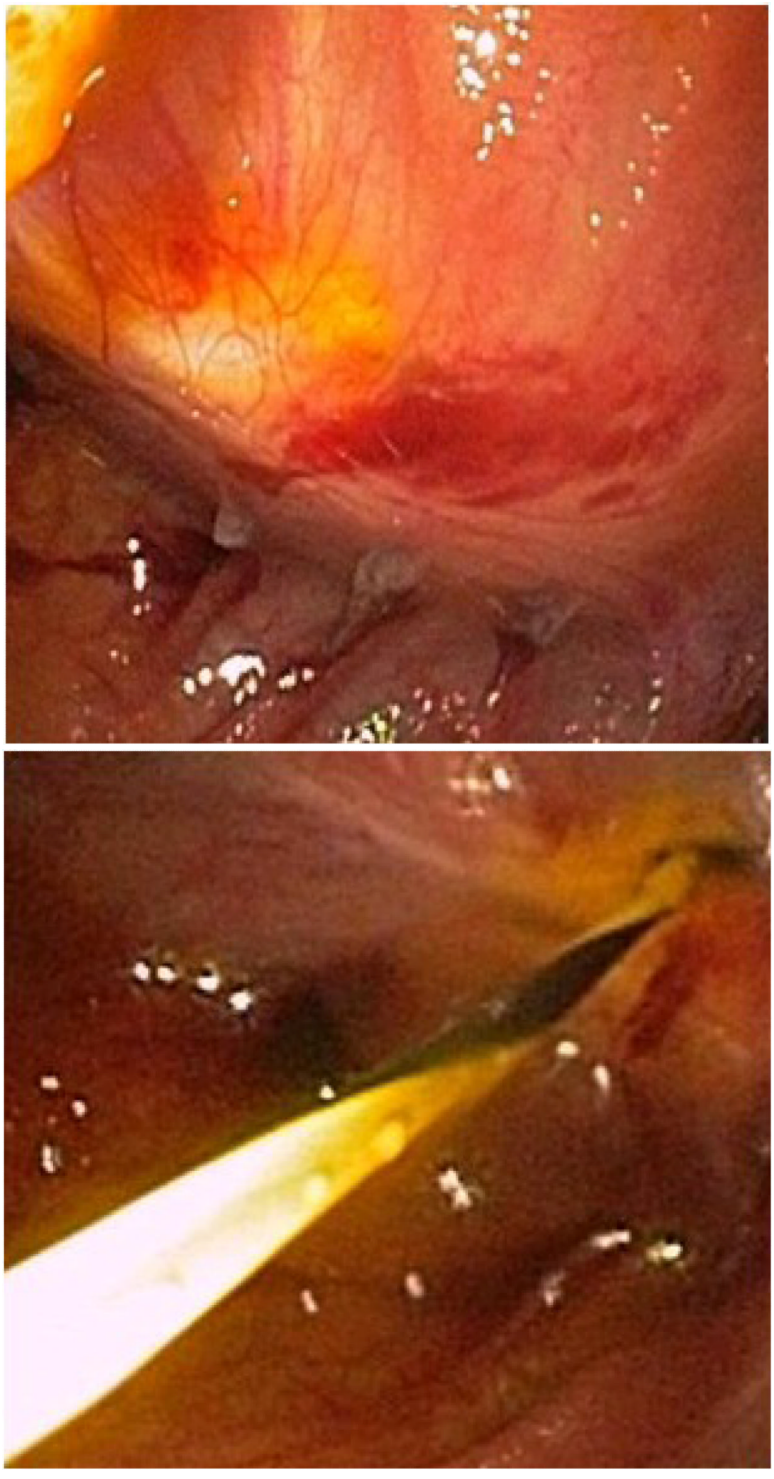


Figure 2.
MRCP image demonstrating diffuse biliary dilatation with left lobe hypertrophy.



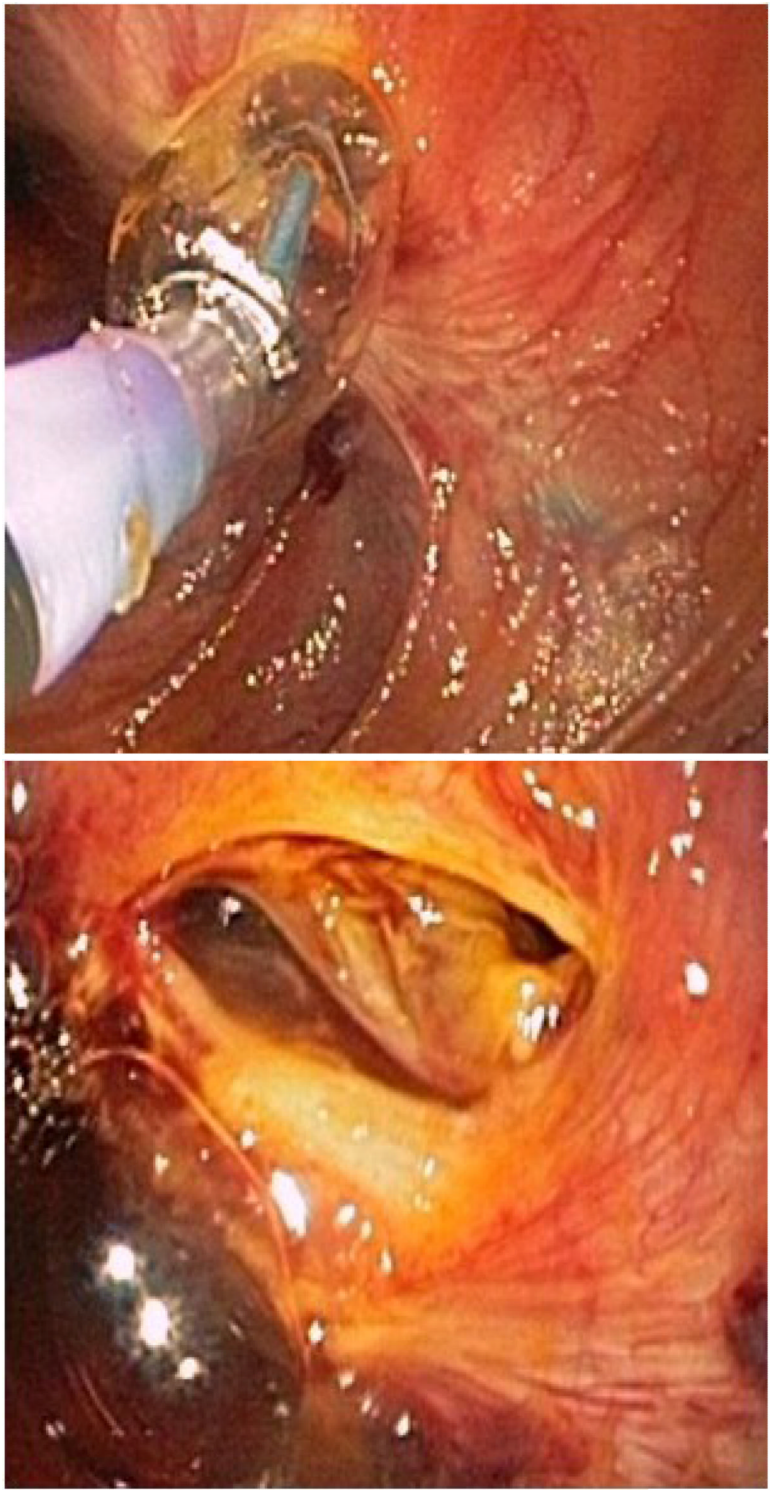
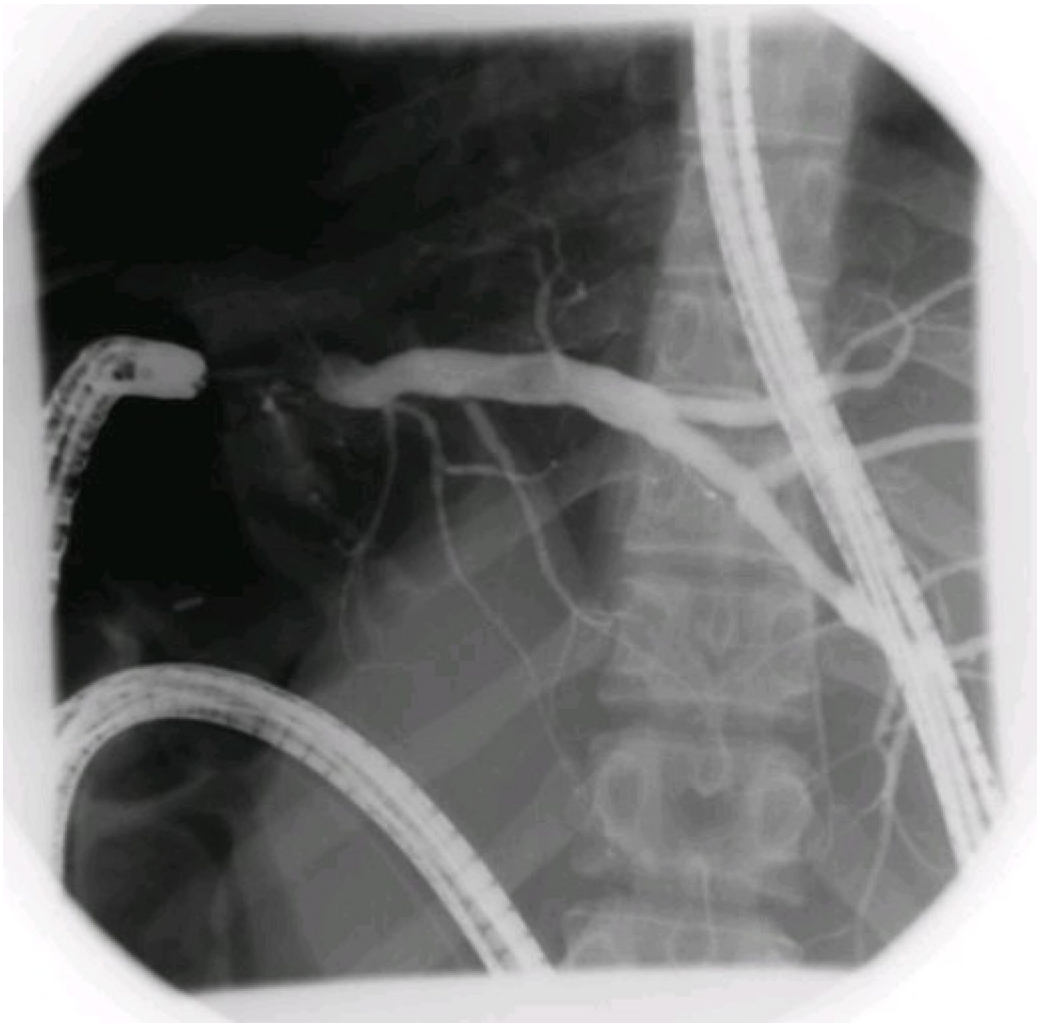


Figure 3.
Endoscopic view of the portoenterostomy. A. Pooling of bile without a readily identifiable biliary orifice. B. Placement of a guidewire across the portoenterostomy. C. Balloon dilation of the orifice to 10 mm. D. Widely patent anastomosis following dilation.



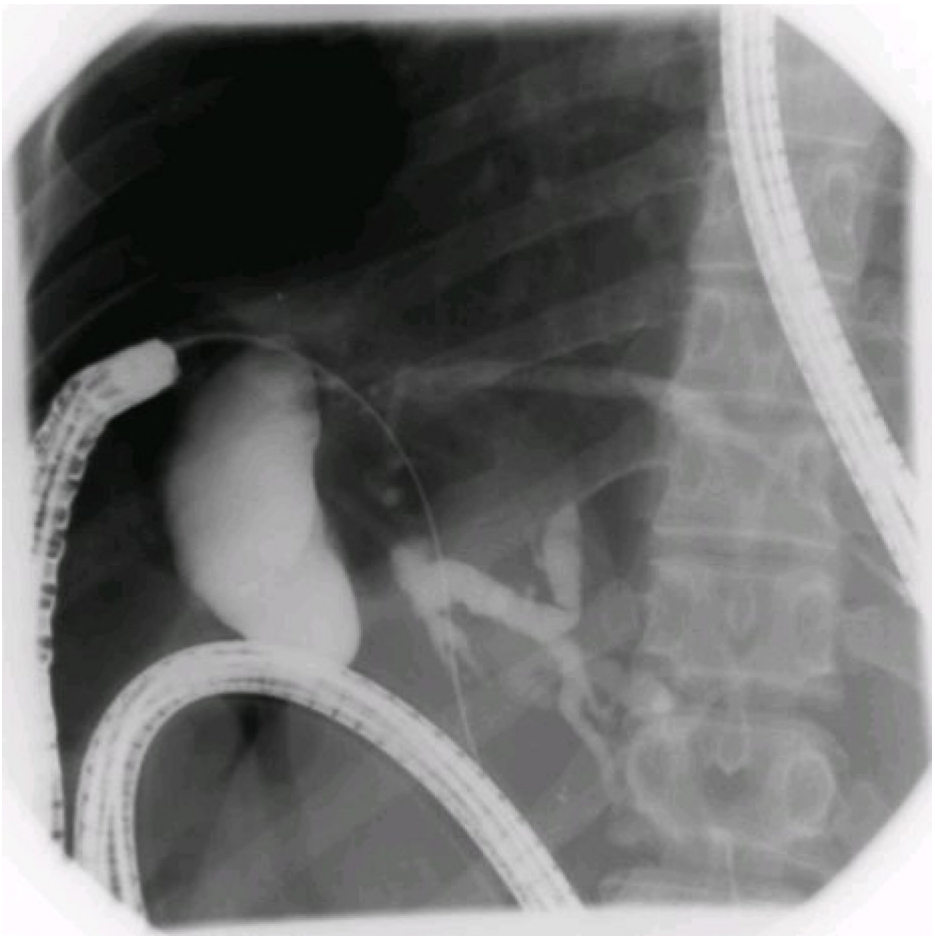


Figure 4.
Fluoroscopic image demonstrating dilatation of the superior duct (A) and inferior duct (B).