Journal of Pediatric Surgery (2012) 47, E1–E5

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Single-balloon enteroscopy-assisted endoscopic retrograde cholangiopancreatography for treatment of cholangitis in a patient with a Kasai portoenterostomy ☆

Eric S. Orman*, C. Brock Miller, Ian S. Grimm, A. Sidney Barritt IV

Department of Medicine, Division of Gastroenterology and Hepatology, University of North Carolina, Campus Box 7080, Chapel Hill, NC 27599-7080, USA

Received 16 August 2011; revised 9 December 2011; accepted 9 December 2011

Key words:
ERCP;
Kasai;
Portoenterostomy;
Single-balloon enteroscopy

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 endoscopic retrograde cholangiopancreatography has not been previously described. Here, we report a patient with recurrent cholangitis after a Kasai procedure and an anastomotic stricture successfully treated with single-balloon enteroscopy-assisted endoscopic retrograde cholangiopancreatography. 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.

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 [1]. The surgery is performed by creation of an anastomosis between a Roux-en-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 most patients developing cholangitis and liver failure and often requiring liver transplantation [1-3]. Previously reported treatment of recurrent biliary obstruction and cholangitis has been limited to antibiotics, corticosteroids, percutaneous transhepatic cholangiography (PTC), and surgical revision [4-6]. 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 anatomy. However, recent advances in deep enteroscopy have allowed endoscopic access to the bile ducts of patients with Roux-en-Y anatomy after gastric bypass and liver transplant [7-9]. Single-balloon enteroscopy, which is commonly used to

Abbreviations: PTC, percutaneous transhepatic cholangiography; ERCP, endoscopic retrograde cholangiopancreatography; MRCP, magnetic resonance cholangiopancreatography.

☆ Funding sources: This work was supported, in part, by the National Institutes of Health, 1KL2-RR025746-03 (Barritt) and T32 DK07634 (Orman).

* Corresponding author. Tel.: +1 919 966 2516; fax: +1 919 966 1700.
E-mail address: eorman@unch.unc.edu (E.S. Orman).

0022-3468/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.12.017

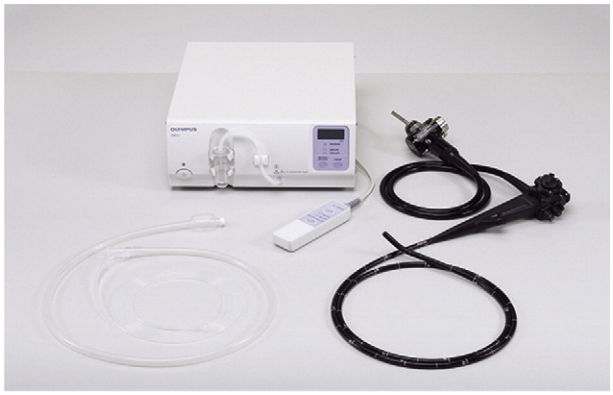

Fig. 1 Single-balloon enteroscopy system. Endoscope pictured on the right and balloon-fitted overtube on the left. The overtube balloon is inflated and retracted to pleat the small bowel, and the endoscope is advanced. This technique is repeated multiple times to achieve deep intubation of the small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

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 (Fig. 1). The enteroscope itself is not designed specifically for ERCP, but devices designed for biliary cannulation and intervention can be passed through the enteroscope. Endoscopic retrograde cholangiopancreatography 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.

1. Case report

A 19-year-old woman was admitted to the hospital for her fourth episode of cholangitis in 4 years. She was diagnosed in utero as having 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 years, 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 years, MRCP was performed showing marked biliary dilatation with hypertrophy of the left hepatic lobe (Fig. 2). Because of 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 (Fig. 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 (Fig. 3B). Contrast injection via the catheter confirmed positioning within a bile duct (Fig. 4A). The orifice was balloon dilated

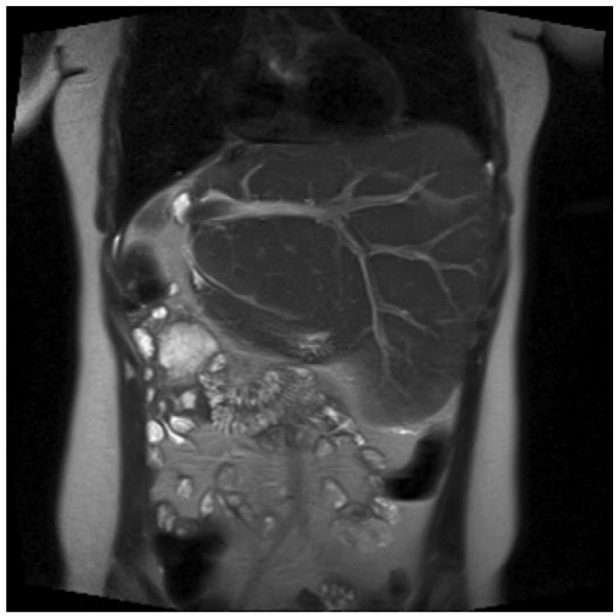

Fig. 2 Magnetic resonance cholangiopancreatography image demonstrating diffuse biliary dilatation with left lobe hypertrophy.