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Percutaneous transhepatic cholangioplasty to treat multiple intrahepatic biliary strictures after hepatoportoenterostomy

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Abstract

In children with biliary atresia (BA), hepatoportoenterostomy (HP) is recommended to improve bile flow. Biliary strictures are known potential complications following HP which can again impair bile flow often leading to biliary cirrhosis and liver transplantation. In patients who are status post HP and have biliary strictures, non-surgical therapeutic options such as endoscopic dilation can pose technical difficulties due to altered anatomy. Percutaneous transhepatic cholangiography with cholangioplasty (PTC-C) is a valuable tool for obstructive cholangiopathies, but to our knowledge this has not been previously demonstrated to be successful in patients with multiple intrahepatic biliary strictures status post HP. Herein, we present three patients status post HP who presented with multiple intrahepatic biliary strictures and underwent successful PTC-C.

Introduction

In children with inadequate biliary drainage, such as in biliary atresia (BA), hepatoportoenterostomy (HP) can be performed to improve bile flow. Biliary strictures are known potential complications following HP which can again impair bile flow often leading to biliary cirrhosis and liver transplantation (1). In patients with BA, despite surgical intervention with HP, progressive liver fibrosis and need for liver transplantation ultimately occurs in 70–80% of children and at least half require liver transplantation by 2 years of age (2–4). Systematic reviews have found that majority of patients status post HP surviving with their native liver develop liver related complications most commonly recurrent cholangitis and intrahepatic biliary strictures (5,6). Liver transplantation can be associated with significant morbidity and mortality, therefore any therapeutic intervention to maximize successful HP would be valuable (7).

Non-surgical therapeutic options, such as endoscopic retrograde cholangio-pancreatography (ERCP), to relieve biliary obstruction due to intrahepatic biliary strictures are technically challenging status post HP due to *roux-en-Y* anatomy (8). PTC-C can also be challenging and has had varied success reported in the literature for intrahepatic biliary strictures post

HP. Kawarasaki et al, categorized intrahepatic biliary cysts as type A non-communicating solitary, type B communicating solitary and type C multi-cystic dilatations. This multi-cystic phenotype has been associated with repeated episodes of cholangitis, a poorer prognosis and inability for permanent alleviation of biliary obstruction via PTC-C or HP revision (9–11). Herein, we present three patients status post HP who presented with multi-cystic intrahepatic biliary strictures and recurrent cholangitis who underwent PTC-C who represent a spectrum of treatment success with majority of patients still possessing their native liver.

Cases

Case 1 is a 24 year old female with BA who presented at 18 years of age with acute cholangitis and eventually developed recurrent cholangitis. She was on prophylactic antibiotics for the first two years post HP. She underwent her first PTC-C at age 21 years. PTC revealed an irregular intrahepatic ductal system with multiple irregular branches coursing down toward the porta hepatis. No right, left or common hepatic ductal system could be identified. (See Figure 1). She required 3 PTC-C procedures in total within 1 week to achieve successful drainage of her biliary system. She had not had any further intervention required, however recently she did have her first episode of cholangitis at 3 years post PTC-C. Other than the recent episode of cholangitis, her AST, ALT, bilirubin and liver function have remained within normal limits during the follow-up period.

Case 2 is a 25 year old female with type IV choledochal cyst status post choledochal cyst excision hepatoportoenterostomy in infancy who presented at age 5 years with cholestasis and eventually developed recurrent cholangitis. She was treated with prophylactic antibiotics during pregnancy, but otherwise did not receive prophylactic antibiotics. She underwent her first PTC-C at age 17 years. PTC revealed multiple chronic biliary strictures from the *roux-en-Y* anastomosis. (See Figure 1). Over the last 8 years, she has had 4 episodes of ascending cholangitis, and has continued to require a biliary drain on account of recurrent stricture formation. She has received about 50 PTC-C procedures in total. Her AST, ALT and direct bilirubin have waxed and waned, but her liver function remains intact.

Case 3 is an 18 year old female with BA who presented for a third opinion after years of recurrent cholangitis. She was on chronic antibiotic prophylaxis. She underwent her first PTC-C at age 17 years. Initial PTC revealed multiple irregular bile ducts (See Figure 1). After her first PTC-C, she developed a fever and was diagnosed with acute cholangitis. She was treated with antibiotics and repeat PTC-C which included balloon dilation of a narrowed *roux* limb, which was previously not identified by other imaging modalities, as well as placement of a right and left biliary drain. After balloon dilation and drainage catheter placement she did have drainage of contrast into the small bowel (See Figure 1). She underwent 4 PTC-C procedures in total. She had initial improvement in cholestasis following PTC-C but ultimately developed subsequent episodes of cholangitis. Given that in addition to the multi-cystic intrahepatic dilations, this patient also had a discrete stricture at the *roux* limb, treatment options including HP revision to address the stricture and liver transplantation were discussed between the medical and surgical teams and the family. Given the normal synthetic liver function our team recommended stricturoplasty but the family elected to seek the option of liver transplantation at another center.

Additionally, all 3 patients also had evidence of normal liver synthetic function throughout their follow-up period, as evidenced by normal glucose, albumin and INR; with the exception of an isolated elevated INR of 1.4 in Case 3 prior to her liver transplantation. As the family opted to undergo liver transplantation at a transplant center closer to their home, it is unclear if this isolated INR, in the setting of prior normal INR values, was due to fat soluble vitamin deficiency. Furthermore, none of the 3 patients had evidence of portal hypertension as evidenced by both normal platelet counts as well as normal hepatopedal flow on liver ultrasounds with doppler. All 3 patients had initial improvement in cholestasis following PTC-C, however this improvement was not long lasting in all of the patients (See Figure 2).

Discussion

In summary, all 3 patients had a history of HP in infancy and later developed recurrent cholangitis and multiple intrahepatic biliary strictures. All patients underwent PTC-C demonstrating a complex multi-cystic pattern of dilation of their intrahepatic biliary system. Although, the number of procedures required varied between patients, all 3 patients had initial improvement in cholestasis following PTC-C (See Figure 2). Two of the three patients had a significantly reduced frequency of cholangitis over a relatively long minimum follow-up period of 3 years. Case 3 had an episode of acute cholangitis following her initial PTC-C which was treated with antibiotics and symptoms resolved once biliary flow was re-established with subsequent PTC-C. There were no other adverse events.

In a relatively recent review of 12 patients with BA status post HP with multiple complicated cystic dilations, PTC-C was shown to be initially successful for cyst decompression but all patients had recurrence of intrahepatic biliary cysts and cholangitis even despite repeated PTC-C (12). The authors concluded that patients with multiple complicated cystic dilations ultimately will require liver transplantation (12). The cases we present possess the same multiple cystic dilation phenotype, but two of the three still possess their native liver without signs of liver failure or portal hypertension at this time. Of note, the patients in this series have survived to adolescence or early adulthood with their native liver and therefore may represent a subset of patients with this multi-cystic phenotype which have milder disease amenable to treatment options other than liver transplantation.

We demonstrate that PTC-C is a safe procedure that can be effective in this difficult patient population with multiple intrahepatic biliary strictures status post HP. However, successful PTC-C in this clinical setting is challenging and is dependent on the expertise of the interventional radiologist and therefore this may limit its widespread applicability. Additionally, case 2 requires permanent biliary drainage, which although PTC-C has allowed this patient to forego liver transplantation, there are potential long term implications including quality of life and risk of cholangiocarcinoma or hepatocellular carcinoma. Despite having a permanent biliary drain, case 2 has successfully carried a child through pregnancy and is a mother of a healthy child.

To our knowledge this is the first successful demonstration of PTC-C in this particular subset of patients. Moreover, in the hands of a skilled interventional radiologist PTC-C may

be either a temporizing measure or possibly a therapeutic option that can decrease the overall need for liver transplantation, but further prospective studies are needed.

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Fig.1.

Fluoroscopic images shown pre and post PTC-C for the 3 cases discussed. a) Irregular intrahepatic ductal system with multiple irregular branches coursing down toward the porta hepatis. b) Early filling of contrast in the small bowel via the intrahepatic ducts c) An abnormal ductal system, with the appearance of multiple strictures and areas of dilatation. d) Flow of contrast through the strictures into small bowel e) An irregular network of bile ducts with areas of both stricturing and beading. f) Focal narrowing at the *roux* limb, but with contrast flowing into the small bowel.

Fig.2.

Line graph demonstrating direct bilirubin values for the 3 cases discussed. Time T=0 indicates the time of the first PTC-C procedure.