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Case Report

Successful percutaneous treatment of biliary stenosis after living donor liver transplantation in a child

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ABSTRACT

We report the case of a 16-year-old boy with primary sclerosing cholangitis associated with inflammatory bowel disease who was initially treated and controlled pharmacologically. He underwent living donor liver transplantation (LDLT) after he developed progressive biliary tract abnormalities and portal hypertension accompanied by recurrent bile duct infections. Two months following LDLT, the hepaticojejunostomy anastomosis became occluded and it was treated surgically. Few weeks later, an increase in drain production persisted without focus; therefore, further diagnostic tests were conducted which proved the recurrence of biliary cast. Under sonographic guidance, external drainage of bile was carried out by percutaneous transhepatic cholangiography and drainage. In total, our patient underwent 5 interventions under general anesthesia and clinically, our patient's general condition improved, and he gained weight. Minimally invasive procedures such as percutaneous transhepatic cholangiography and drainage and balloon dilation are safe and effective, but may require several attempts before being successful.

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Introduction

Liver transplantation has been successfully performed in pediatric patients with end-stage liver disease. The most common causes of liver failure in pediatrics are (but not limited to) primary sclerosing cholangitis (PSC) and biliary atresia. A significant advance in transplantation has been the use of living donor and split-liver grafts, which result in a

reduced morbidity and mortality. Clinically, transplantation requires a vascular reunion of the portal vein, hepatic artery, and hepatic veins between the graft and host. Moreover, continuity of the bile duct with the gastrointestinal tract must be established, for example, via hepaticojejunostomy. Vascular anatomy, sufficient volume for the metabolic demands, and biliary drainage are regarded thus far as the technical challenges. Biliary strictures occur in up to 1/3 of pediatric patients following liver transplantation, and these are often

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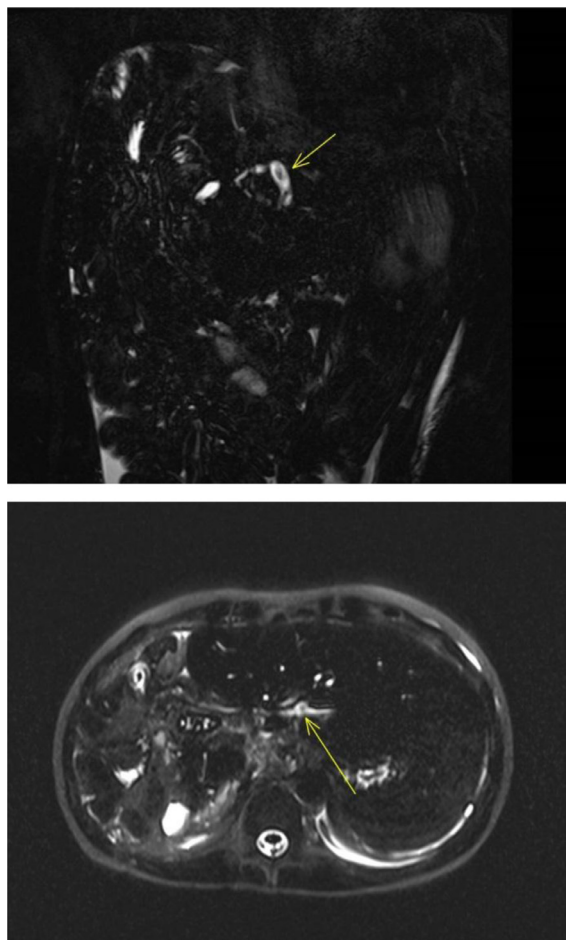


Fig. 1 – MRCP coronal (A) and axial (B) images show the dilation of the intrahepatic duct, in particular segment 2/3. MRCP, magnetic resonance cholangiopancreatography.

managed by interventional radiologic techniques. The main advantage of interventional radiology techniques is the ability to treat common complications such as biliary strictures percutaneously via a minimally invasive method; therefore, the need for surgery will be minimized with the aim of restoring the function of the transplanted liver. As a result, the interventional radiologist has become one of the most important members in the multidisciplinary transplantation team. The most common liver transplantation method is an orthotopic liver transplantation where the donor organ is placed in the same anatomic location as the original. The technique described in this paper applies to living related. In this paper, we report the case of a 16-year-old boy with PSC associated with inflammatory bowel disease who underwent a liver transplantation.

Case presentation

The patient is a 16-year-old boy with known Crohn's disease, PSC, choledochocystolithiasis, and irregular bile ducts.

He presented to his general pediatrician with chronic abdominal pain for 6 months accompanied by diarrhea, reduced appetite, and malaise since these complaints were getting worse in the course of time. On examination, the liver was palpable 2 cm below the rib cage. Furthermore, the pediatrician excluded celiac disease and infections (in particular hepatitis B, Epstein–Barr virus, and Cytomegalovirus). The ultrasound of the upper quadrant of the abdomen showed anatomic abnormalities in the bile ducts. Moreover, the patient was sent to the department of gastroenterology at the University Medical Center for further diagnosis. The pediatric gastroenterologist performed a colonoscopy, gastroscopy, and histology of the liver from which the diagnosis of PSC associated with Crohn's disease was confirmed. Over a period of time, the patient was treated conservatively. Few months later, the patient underwent an LDLT. Following the transplantation, his condition is improved as well as the liver function tests. On day 14 post-operative, the hepaticojejunostomy anastomosis became occluded which led to progressive intrahepatic cholestasis and abdominal complaints and as a result were treated surgically. On day 38, an increase in drain production persisted without focus. Therefore, an ultrasound and magnetic resonance cholangiopancreatography were performed which showed dilated intrahepatic bile ducts and fluid collection in the central intrahepatic duct, in particular segments 2 and 3 which confirmed the recurrence of biliary cast (Fig. 1).

It was advised to place a percutaneous transhepatic cholangiography and drainage (PTCD) under ultrasound guidance (Fig. 2). During PTCD, a thin needle is inserted percutaneously and advanced through liver tissue under ultrasound guidance until a bile duct is entered. A contrast material is used to outline the bile duct system. When the correct position of the needle is radiographically confirmed, a catheter is inserted to allow the bile to drain either into a small pouch attached outside the body or into the small intestine. During the procedure, a low-grade stenosis of the anastomosis was seen. This was not treated with percutaneous transluminal angioplasty (PTA) because of recent anastomosis. After PTCD, the patient's general condition improved. The patient was advised to return within 5–6 weeks for a follow-up and PTA if needed.

Between May 2018 and October 2018, 5 attempts were made to restore the bile flow percutaneously as surgical options were not necessary, unless unavoidable. If needed, the occluded bile ducts were dilated by means of PTA (Fig. 3). This effectively restored the bile drainage. A few months following the last PTCD, the patient was well, and the transplanted liver had gained its function as expressed by stable liver parameters (aspartate transaminase 61U/L, alanine aminotransferase 53U/L, gamma-GT 213U/L, total bilirubin 15 $\mu\text{mol/L}$).

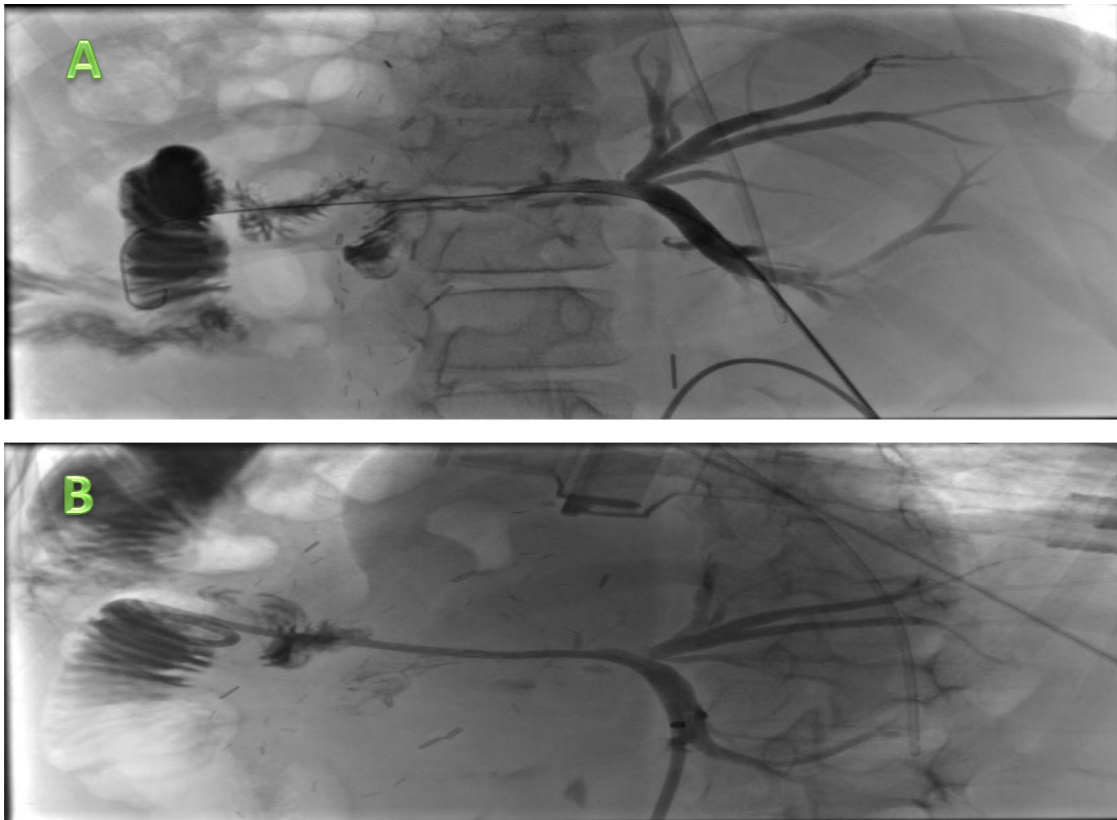


Fig. 2 – Cholangiography of the liver transplant recipient. (A) shows irregular form of the anastomosis without signs of sludge. In consultation with the pediatric gastroenterologist, it is decided to place a biliary drain (8.5 Fr) without PTA as shown in (B).

PTA, percutaneous transluminal angioplasty.

Discussion

Biliary strictures are one of the most common complications following a living-related liver transplantation in pediatric patients with PSC. Biliary strictures in the anastomosis have been reported in up to 40% of recipients [1]. Nonsurgical intervention of biliary complications is set to be the preferred diagnostic and therapeutic option, as surgical intervention is associated with significant postoperative morbidity [2]. Minimally invasive procedures such as PTA and PTCD are proven to be safe and effective, though several attempts may be required in order to reach a successful outcome [3]. A study conducted by Moreira et al. showed that repeated re-interventions increased the complication rate [4]. When PTCD fails, the follow-up usually includes surgical revision of the transplanted liver. Patients who are younger at the time of transplantation are at a higher risk of developing biliary strictures. If biliary strictures occur within 30 days following the liver transplantation, Karakayali et al. showed that it must be assumed that the reason for this was a problem with the surgical technique [5].

In our case, the patient was exposed to anesthesia 5 times during the minimally invasive procedures. Anesthesia performed in a weak child with quite a moderate condition remains a challenge for therapists. Effective co-operation be-

tween the anesthesia and pediatric intensive care unit is essential during the perioperative period. The nutritional status of the patient has an important role on the peritransplant outcome. Cholestatic liver disease plays a role in fat malabsorption accompanied by caloric deficit as well as fat-soluble vitamins deficiency in which it necessitates an assessment of the intake and supplemental feeding by means of tube or infusion [6]. Moreover, not fully developed immune system in pediatric patients combined with use of immunosuppressive agents increases the risk of infections and complications [7].

As already known, the first PTCD and (PTA if carried out) procedures usually pose the greatest challenge to interventional radiologists due to a number of reasons; the degree of strictures are higher, more fibrotic tissues, narrowing of the ductal lumen and many more [8]. Furthermore, it is usual for the patients to present with cholangitis in which the volume of contrast should be minimized to prevent sepsis [9]. Over the course of the time and when the patient has an internal/external drain in situ, the procedures become less complicated as the route is secured and the strictures have already been dilated for at least once before. Belenky et al. advised that in the case of late biliary strictures, the treatment option should be stent placement which shows long-term results in comparison to drains or balloons [9]. In the study conducted by Cardarelli-Leit et al., they avoided placing stents in pediatric

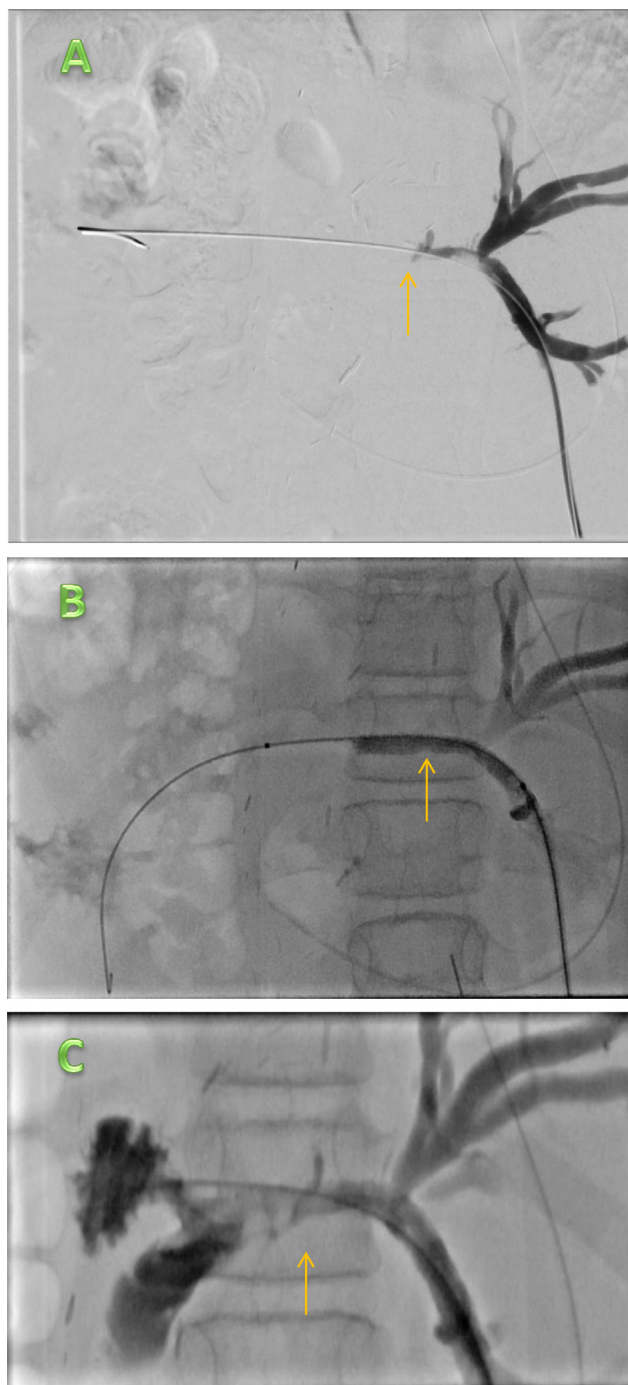


Fig. 3 – Follow-up cholangiogram after 6 weeks following the 1st intervention. (A) shows complete occlusion in the segment 4 branch of the bile duct, promoting ductal dilatation. (B) shows PTA of the whole pathway using different angioplasty balloon catheters. (C) shows the result of sequential cholangioplasty. No evidence of stenosis with respect to the anastomosis. PTA, percutaneous transluminal angioplasty.

patients since they are susceptible to obstruction over time which creates difficulty for those who will have to undergo new surgery in the future [9,10]. The industry has recently introduced a covered self-expandable nitinol stent that can prevent the aforementioned issues. Despite their high cost, the use of such stents in the case of benign biliary strictures in pediatric patients needs further study [9,10].

Conclusion

Biliary strictures are a frequent complication following liver transplantation in pediatric patients. Minimal invasive procedures such as percutaneous drainage placement and balloon dilation are safe and effective but may require several attempts before being successful.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.03.039](https://doi.org/10.1016/j.radcr.2019.03.039).

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