Technical Challenges of Percutaneous Transhepatic Biliary Drainage in Adult Biliary Atresia Patients Post Kasai Portoenterostomy

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Radiology Case. 2024 November; 18(11):16-23 :: DOI: 10.3941/jrcr.5473

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Conflict of Interest / Disclosures

The authors declare no financial or non-financial competing interests related to this study.

Consent

Yes, written informed consent was obtained from all patients involved in this study for the submission and publication of this manuscript.

Conflict of Interest / Human and Animal Rights

This study involved human participants and was conducted in accordance with the ethical standards of the institutional and national research committees. It adhered to the principles outlined in the Helsinki Declaration of 1975, as revised in 2000.

ABSTRACT

Biliary atresia (BA) is a severe hepatobiliary condition requiring Kasai portoenterostomy (KP) during infancy to facilitate biliary flow. However, post-KP, patients frequently develop complications such as recurrent cholangitis, biliary stricture, and cystic intrahepatic dilatation. These complications often progress to biliary cirrhosis and end-stage liver disease, necessitating liver transplantation.

In this context, we explore the application of Percutaneous Transhepatic Biliary Drainage (PTBD) for treating biliary complications in post-KP BA patients. PTBD in these cases is technically challenging due to the presence of biliary strictures and intrahepatic bile duct dilatation, and its effectiveness has been a subject of debate.

This paper presents two technically challenge cases where we treated adult BA patients who developed late-onset biliary strictures following successful KP. Both of the required first direct puncture of a cystic region of biliary dilatation, followed by successful puncture (one patient was punctured on CT-guidance) and wire cannulation and successful PTBD insertion. Following the procedure, both patients achieved short-term and long-term clinical improvement. The significant findings from these cases include: resolution of acute cholangitis, normalization of liver enzymes, no evidence of biliary cirrhosis in subsequent ultrasound, and removal of both patients from the liver transplantation list.

CASE REPORT

BACKGROUND

Kasai portoenterostomy (KP) is a surgical procedure performed in early infancy for patients with biliary atresia (BA) to re-establish bile flow [1]. Even in patients who achieve jaundice clearance following successful KP, late biliary complications such as recurrent cholangitis and intrahepatic biliary stricture and dilatation may occur . Kawarasaki et al categorized the intrahepatic biliary dilation into three types: type A, non-communicating solitary cyst, type B, communicating solitary cyst, and type C, multi-cystic dilatation [2]. The cystic

dilatation of the intrahepatic biliary system predisposes to a vicious cycle of recurrent cholangitis and progressive strictures and can rapidly lead to liver decompensation requiring liver transplant (LT) [1,3-5]. Interventions to maintain functional KP and delay liver transplantation are of great clinical value.

Despite a few case series demonstrating the percutaneous transhepatic biliary drainage (PTBD) procedure post-KP in pediatric BA patients, its clinical effectiveness varies [6-8]. Studies on PTBD in adult BA patients are rarer. Some reported technical challenges and limited effectiveness due to difficulties in navigating guidewires through intrahepatic bile duct dilatations and strictures [9]. Another study reported the effectiveness of PTBD, but did not reveal technical details of the procedure [10].

In this study, we described a novel technique applied to two adult patients with biliary atresia (BA) post-Kasai portoenterostomy (KP), who presented with recurrent cholangitis and multicystic intrahepatic biliary strictures in their native livers. For both patients, we first targeted and punctured the most prominent cystic dilatation. This initial step was crucial as it provided a detailed cholangiogram, which delineated the anatomical structures of the intrahepatic biliary ducts, precisely located the site of the portoenterostomy anastomosis, and assessed the extent of the strictures. Following this, the intrahepatic duct was selectively punctured. This strategic choice facilitated effective cholangioplasty to address the stenosis at the portoenterostomy anastomosis site. Additionally, we underscore the importance of utilizing CT guidance in a case where fluoroscopy alone does not sufficiently reveal the biliary ducts' structure and alignment, thus ensuring a more accurate and safer intervention.

CASES

Case 1

Case 1 is a 29 year-old Chinese female diagnosed with BA and underwent KP during her infancy. She presented with recurrent cholangitis and underwent first percutaneous cholangiography and cholangioplasty at the age of 24 years old. MRI showed diffuse dilatation of the intrahepatic biliary ducts in both hepatic lobes and the dilated biliary ducts demonstrated beaded appearance (Figure 1D,1E). Two prior PTBD attempts from the right duct were unsuccessful. On this occasion, the right hepatic duct was punctured initially by a 22G Chiba needle (Cook Medical, Indiana, United States). Contrast was injected into the intrahepatic ducts, revealing multicystic intrahepatic biliary duct dilatations (type C), however, the guidewire was unable to transverse the portoenterostomy to reach the duodenum as with previous attempts. During the repeat attempts, the puncture access was lost, and the track could not be visualized. Subsequently, decision was made to puncture the left hepatic duct with a 21G Accustick needle (Boston Scientific, Massachusetts, United States). It was then when it became clear that the anastomosis involved the left duct only. However, there was still difficulty in passing the wire into the intrahepatic ducts, after several attempts decision was made to switch to CT guidance. On CT scan, a relatively good sized cystic dilatation of the duct that could be selected for puncture, which was successful. Our choice of wire for these tortuous ducts is the 0.018" Glidewire (Terumo Medical Corporation, New Jersey, United States). The portoenterostomy was successfully crossed and dilated with a 6 x 20 mm cutting balloon (Boston Scientific, Massachusetts, United States) and 8 x 40 mm sterling balloon (Boston Scientific, Massachusetts, United States), followed by the insertion of an 8 F PTBD tube. Over the next 10 months, she underwent 5 additional interventions for biliary stricture dilatation and PTBD tube changes. Following these interventions, the PTBD was removed. The patient was then followed up for 54 months, during which she experienced no further cholangitis episodes, maintained normal liver function and liver enzymes, and showed no signs of biliary duct dilatation or liver cirrhosis on interval ultrasound scans. She remains off the liver transplantation list.

Case 2

Case 2 is a 21 year-old Chinese male, who underwent KP during his infancy. He presented at 19 years old with recurrent cholangitis and severe jaundice. MRI showed focal cystic dilatation of the intrahepatic biliary duct in the region of the hepatoportoenterostomy anastomosis (Figure 2C,2D). The central cystic dilatation was accessed using a 22G Chiba needle (Cook Medical, Indiana, United States). Contrast was injected into the intrahepatic ducts, revealing a focal cystic dilatation communicating with the intestinal loop at the portoenterostomy anastomosis site, and mild intrahepatic bile duct dilation (type B). Then one of the right hepatic ducts was then punctured using a 21G Accustick needle (Boston Scientific, Massachusetts, United States) under fluoroscopy. After crossing of the right hepatic duct stricture, cholangioplasty was performed using a 4 x 40 mm Powerflex PRO balloon (Cordis, Florida, United States) and an 8F Skater biliary catheter (Argon Medical, Texas, United States) was inserted for internal and external drainage and below the obstruction. The distal catheter loop was positioned within the duodenum. Over the following 9 months, he underwent 7 additional interventions including PTBD tube change and stricture dilatation. Eventually, PTBD was removed. At a 42-month follow-up, the patient had no further cholangitis episodes, normal liver function and liver enzymes, and interval ultrasound scans showed no signs of biliary duct dilatation or liver cirrhosis. He is also not on the liver transplantation list.

DISCUSSION

PTBD in post KP patients can be challenging. Some patients have gracile intrahepatic ducts (as with patient 2) and some patients have chronic multifocal stenosis (as with patient 1). Both PTBDs were successful by first puncturing a cystic region followed by puncturing a favourable intrahepatic duct. Uniquely in patient 1, we found that the portoenterostomy involved only the left duct and CT guidance was used to find and target the most favourable intrahepatic duct to gain access. Both patients demonstrated favorable long-term clinical outcomes. Notably, they remained free from cholangitis throughout the follow-up period. Their liver function tests, including total bilirubin, liver enzymes, and synthetic functions, consistently stayed within

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normal ranges. Additionally, interval ultrasound examinations of the liver revealed no signs of biliary duct dilatation or liver cirrhosis. Remarkably, neither patient is currently listed for liver transplantation, underscoring the effectiveness of the interventions in preserving native liver function.

Weidner et al. reported three cases of successful percutaneous cholangiography and cholangioplasty, yet their publication lacked detailed descriptions of the techniques used [10]. In contrast, Onish et al. identified significant challenges in percutaneous transhepatic cholangiography and cholangioplasty, particularly the difficulties encountered in inserting and navigating a guidewire through the complex landscape of multiple dilatations and strictures within the intrahepatic bile ducts [9].

This case series suggest that percutaneous cholangiography and cholangioplasty are effective for managing patients with multiple intrahepatic dilatations and biliary strictures, avoiding a liver transplant at the prime of their lives and, for female patients, child bearing years. However, the success of these interventions heavily relies on the skill and experience of the interventional radiologist.

Nevertheless, given the limited number of cases and the retrospective nature of this series, further research with a larger cohort is imperative to substantiate these findings.

TEACHING POINT

The management of late complications in patients with biliary atresia (BA) post-Kasai portoenterostomy (KP) presents significant clinical challenges, particularly in adult patients. This case series demonstrates the potential of percutaneous transhepatic biliary drainage (PTBD) as an effective intervention for addressing recurrent cholangitis and intrahepatic biliary strictures. Key teaching points include the importance of precise imaging techniques, such as CT guidance, to accurately identify and target cystic dilatations within the intrahepatic biliary system. Moreover, the strategic selection of puncture sites and the use of appropriate guidewires are critical for navigating complex biliary anatomies. These interventions, when performed by skilled interventional radiologists, can significantly delay the progression to liver transplantation, thereby preserving native liver function and improving patient quality of life. This case series also highlights the necessity for ongoing research and case reporting to further refine these techniques and optimize outcomes for this patient population.

QUESTIONS

1. What is the main goal of the Kasai portoenterostomy (KP) procedure in infants with biliary atresia (BA)?
   1. To remove the gallbladder
   2. To establish bile flow
   3. To prevent liver cirrhosis
   4. To perform liver transplantation
   5. To correct portal hypertension

   Answer: 2. To establish bile flow

2. Which type of intrahepatic biliary dilatation involves multi-cystic dilatation?
   1. Type A
   2. Type B
   3. Type C
   4. Type D
   5. Type E

   Answer: 3. Type C.

3. What diagnostic tool was utilized when fluoroscopy alone was insufficient to reveal the biliary duct structure and alignment in the first case?
   1. Ultrasound
   2. Magnetic Resonance Imaging (MRI)
   3. Computed Tomography (CT) scan
   4. Endoscopic Ultrasound (EUS)
   5. Positron Emission Tomography (PET) scan

   Answer: 3. Computed Tomography (CT) scan

4. What was the primary strategy employed in PTBD to manage the stenosis at the portoenterostomy anastomosis site?
   1. Use of endoscopic retrograde cholangiopancreatography (ERCP)
   2. Placement of a biliary stent
   3. Puncturing a cystic region followed by cholangioplasty
   4. Administration of oral antibiotics
   5. Use of radiofrequency ablation

   Answer: 3. Puncturing a cystic region followed by cholangioplasty

5. What was the long-term outcome for both patients following the PTBD?
   1. Both required immediate liver transplantation
   2. Both developed chronic liver disease
   3. Both experienced no further episodes of cholangitis and maintained normal liver function
   4. Both showed increased liver enzyme levels and signs of biliary duct dilatation
   5. Both patients required additional surgical interventions for bile duct reconstruction

   Answer: 3. Both experienced no further episodes of cholangitis and maintained normal liver function

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FIGURES

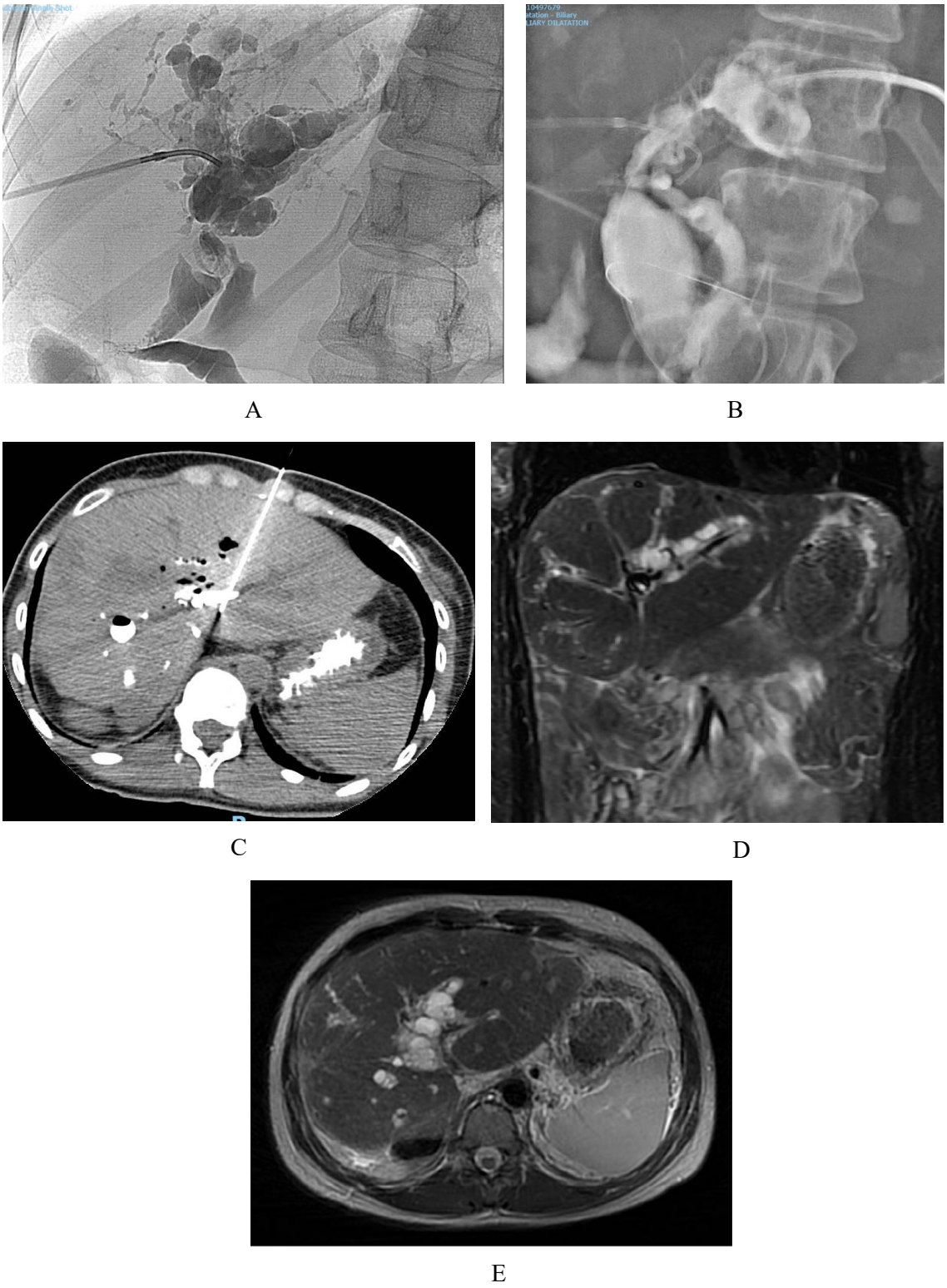

Figure 1 (case 1): A: Cholangiogram from right hepatic duct puncture showing bilateral and multiple stenoses and strictures (type C). Communication with the bowel is not clearly seen. B: Cholangiogram from left hepatic duction showing communication with bowel (path of wire). C: intra procedural CT showing the most suitable area of puncture in the left duct and targeting on CT. D and E: T2 weighted MRI showing diffuse dilatation of the intrahepatic biliary ducts in both hepatic lobes and the dilated biliary ducts demonstrated beaded appearance.

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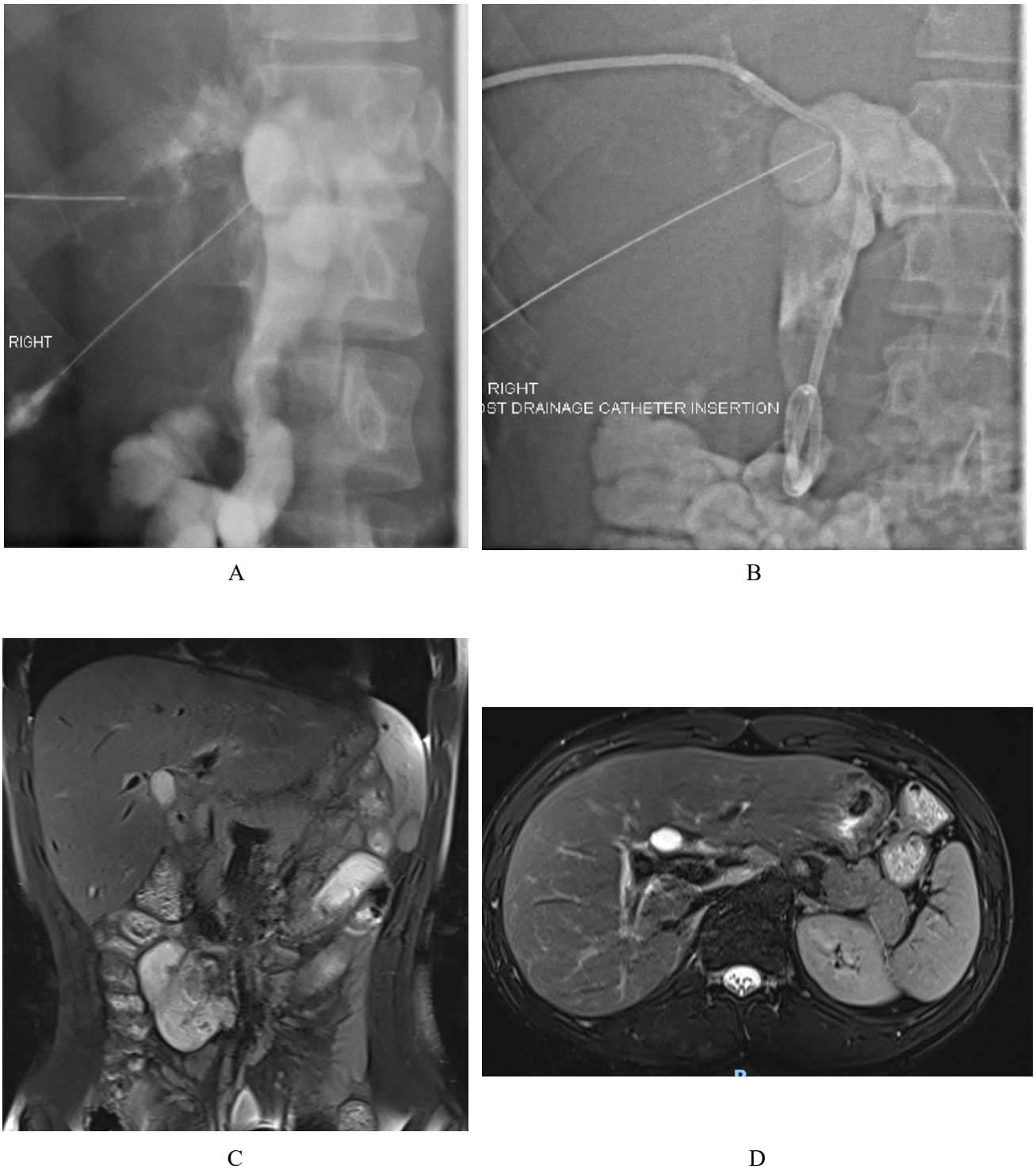

Figure 2 (case 2) A: Cholangiogram obtained from puncturing the solitary cystic lesion (type B) with another needle targeting the gracile intrahepatic ducts. B: Successful insertion of the a biliary drain. C and D: focal cystic dilatation of the intrahepatic biliary duct in the region of the hepatoportoenterostomy anastomosis.

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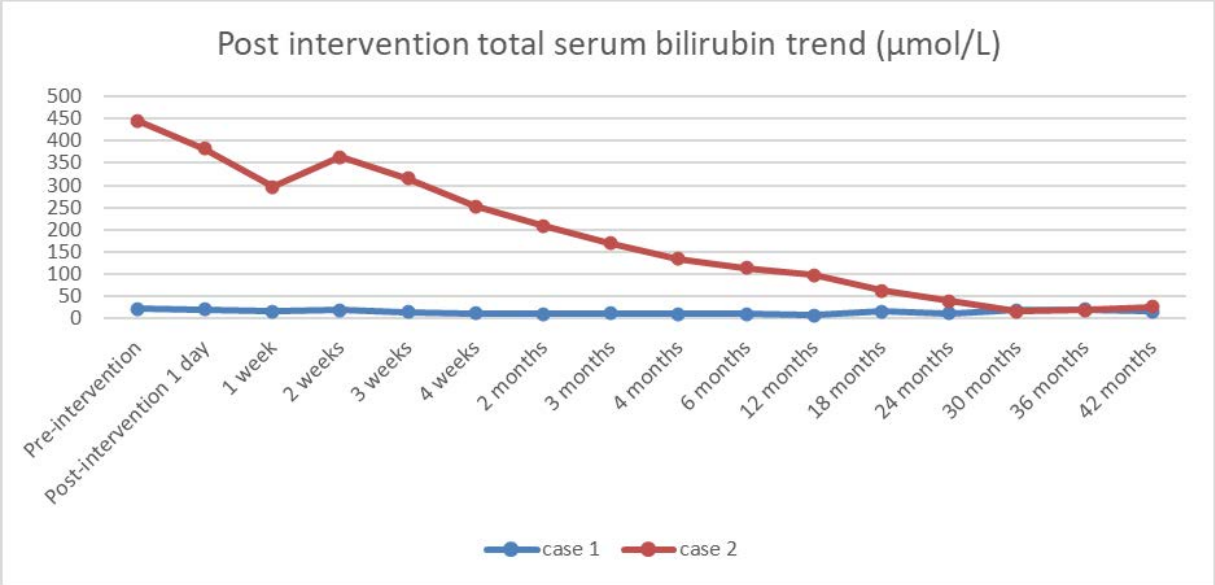

Figure 3: Serum biliary trend pre- and post-intervention

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KEYWORDS

Biliary atresia, kasai portoenterostomy.

ABBREVIATIONS

BA = Biliary Atresia
KP = Kasai Portoenterostomy
LT = Liver Transplant
PTBD = Percutaneous Transhepatic Biliary Drainage
MRI = Magnetic Resonance Imaging
CT = Computed Tomography
ERCP = Endoscopic Retrograde Cholangiopancreatography
EUS = Endoscopic Ultrasound
PET = Positron Emission Tomography

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