Conjugated hyperbilirubinemia represents a diagnostic challenge in the newborn period. Obstruction of the extrahepatic bile ducts is the most common cause of conjugated hyperbilirubinemia [1]. Although it can be caused by a large number of disorders, biliary atresia and neonatal hepatitis account for most of the cases. A rare cause is obstruction of the common bile duct by bile sludge known as bile-plug syndrome (inspissated bile syndrome). The treatment of this condition is generally surgical. Some minimally invasive treatment modalities, such as percutaneous transhepatic cholangiography with contemporary saline lavage of the biliary tree, and laparoscopic aided cholecystostomy for bile duct lavage have also been described in the literature [2,3]. Here, we present a new minimally invasive treatment modality for bile-plug syndrome. © 2011 Elsevier Inc. All rights reserved.

1. Case report

A 5-month-old male infant presented with complaint of widespread papulopustular eruption that had progressed with periods of exacerbation since 1 month of age. The patient’s vital signs were normal. On physical examination, the patient had also a palpable liver 2 cm below the right costal margin. Laboratory tests showed anemia (hemoglobin, 9.1 g/dL), leukocytosis (white blood cell, 22500/mm³), and elevated $\gamma$-glutamyltransferase (GGT, 869 U/L) and alkaline phosphatase (ALP, 545 U/L) levels with normal alanine aminotransferase (ALT), aspartate aminotransferase (AST), and bilirubin levels. Serum cytomegalovirus (CMV) polymerase chain reaction revealed 2860 copies per mL. Abdominal ultrasonography (USG) showed a large and heterogeneous liver with mildly dilated intrahepatic biliary tract and a normal gallbladder.

On the fifth day of hospitalization, mild clinical jaundice was diagnosed on the basis of yellowed sclera. Serum
Bilirubin concentration was 4.3 mg/dL with 2.9 mg/dL direct reacting. The patient had also acholic stools. Abdominal computerized tomography revealed dilatation of the intrahepatic biliary tracts and common hepatic duct and obstruction of common bile duct. Tru cut needle biopsy of the liver showed bridging fibrosis and cholangitis. Medical treatment was started with ursodeoxycholic acid (20 mg/kg per day), cefotaxime, and ganciclovir.

The patient’s liver function tests (LFT) on the 20th day of hospitalization were as follows: ALP 625 U/L (normal up to 120 U/L), GGT 1137 U/L (normal up to 38 U/L), AST 115 U/L (normal up to 31 U/L), ALT 25 U/L (normal up to 34 U/L), conjugated bilirubin 2.94 mg/dL (normal up to 0.2 mg/dL), and total bilirubin 4.65 mg/dL (normal up to 1.2 mg/dL). Ultrasonography-guided percutaneous transhepatic transcholecystic cholangiography was performed under sedation and local anesthesia using a 21-gauge needle, which was inserted from a right anterolateral approach, and showed dilated intrahepatic biliary tract and main hepatic duct and obstructed common bile duct (Fig. 1). A 6F pigtail loop catheter was inserted into the gallbladder for biliary drainage using Seldinger technique under fluoroscopy. The catheter was sutured to the skin, placed on gravity drainage, and then irrigated daily with 5 mL of sterile saline to avoid occlusion. The patient’s LFT were decreased within 2 weeks. Abdominal USG showed normal intrahepatic biliary tracts and main hepatic duct. Transcatheter cholangiography showed continued obstruction of the common bile duct (Fig. 2). A guide wire was inserted through the cholecystostomy catheter under sedation, and common bile duct obstruction was passed using 0.018 in and 0.035 in hydrophilic guide wires under fluoroscopic guidance (Fig. 3); it was dilated by 7 mm/40 mm balloon dilatation catheter (Fig. 4). After this successful intervention, contrast medium passed into the duodenum was observed (Fig. 5).

Fig. 1 Percutaneous transhepatic transcholecystic cholangiography showing dilated intrahepatic biliary tract (black arrow), main hepatic duct (white arrow), and obstructed common bile duct (white arrowhead). The asterisk indicates contrast medium leakage around the gallbladder.

Fig. 2 Transcatheter cholangiography showing the common bile duct obstruction (white arrow).
The preoperative medications were continued after the procedure. The patient passed cholic stool 1 day later. Subsequent transcatheter cholangiography carried out 2 weeks later showed good passage to the duodenum without any extravasation; therefore, the catheter was removed. Liver function tests were as follows: AST 61, ALT 52, GGT 460, ALP 973, total bilirubin 1.55, and conjugated bilirubin 0.64. The recovery period was uneventful. He was

![Image](image1.png)

**Fig. 3** The obstruction was opened using guide wires (white arrow).

![Image](image2.png)

**Fig. 4** The common bile duct was dilated by 7 mm/40 mm balloon catheter (white arrow).
discharged 3 weeks later; then, ursodeoxycholic acid was continued for 2 months.

2. Discussion

Bile-plug syndrome is defined as an obstruction of the common bile duct by bile sludge in full-term infants without anatomical abnormalities, or congenital chemical defects of bile [4]. Any condition leading to alteration in bile composition may cause this syndrome. Although the etiology of bile-plug syndrome is not clear, it can develop in association with abnormal composition of bile in cystic fibrosis, hepatocellular damage, prolonged erythroblastic jaundice, or total parenteral nutrition [5-8]. Dunn [6] observed that the conjugated hyperbilirubinemia is probably related to hepatocellular damage and that cholestasis is secondary. Another study showed that CMV infections of the neonates are associated with the development of cholestatic disorder [8]. In our case, hepatocellular damage caused by CMV infection seems to be a logical etiologic factor leading to bile-plug syndrome.

Although spontaneous resolution has been reported in the literature [9], bile-plug syndrome generally requires surgical intervention. Different surgical procedures such as retraction of the bile plug via a duodenostomy and papillotomy, manual propulsion of the bile plug into the duodenum, and intraoperative irrigation of the biliary system with saline or mucolytic agents have been described in the literature [1,10,11]. Mechanical methods of irrigating the biliary tract provide the potential of improving cholestasis by flushing out inspissated secretions within the biliary tract. Wales et al [2] observed that neonates with cholestasis who received percutaneous transhepatic transcholecystic cholangiography for evaluation of conjugated hyperbilirubinemia experienced more rapid resolution of their cholestasis. However, percutaneous cholangiography in an infant is a difficult procedure. Moreover, it is sometimes difficult to obtain a sufficient flushing pressure.

Over the past few decades, percutaneous biliary interventions have showed a great improvement and may be an alternative to surgery. Such interventions generally are less stressful for children than alternative surgical procedures. The most widely used interventions are percutaneous cholangiography, biliary drainage and stenting, and balloon dilatation, which have an important role in the treatment of a variety of biliary tract disorders. The feasibility of these interventions in adults and some children has certainly been demonstrated [12], but the data about their usage in infants have been very limited in the literature. To our knowledge, only 1 case of infant with common bile duct stricture treated by percutaneous biliary drainage, biliary stenting, and balloon dilation has been reported in the literature [13]. Although our case was treated in a similar way, this is the first report showing an infant with bile-plug syndrome can be treated by percutaneous management without the need for surgery.

Percutaneous biliary interventions can lead to some complications, especially in infants. Although we did not encounter any complications in our case, the possible complications related to this procedure are bile leak and peritonitis, hemorrhage, common bile duct or duodenal perforation, especially after the balloon dilatation.

In conclusion, percutaneous treatment of bile-plug syndrome is reported in an infant as a first case in the literature. This treatment modality may be a useful alternative to surgery in infants with bile-plug syndrome.

References

Percutaneous management of bile-plug syndrome