

# Spontaneous Common Bile Duct Perforation in an Infant Diagnosed by Hepatobiliary Radionuclide Imaging: A Case Report

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## ABSTRACT

Spontaneous biliary duct perforation is a rare yet important entity presenting in infants. The usual presentation is jaundice and failure to thrive occurring in a previously healthy infant. The diagnosis is usually the function of hepatobiliary imaging. In this case report Tc<sup>99m</sup>-DISIDA hepatobiliary imaging was an accurate tool to diagnose this surgically correctable disease.

**Key words:** Common Bile Duct Perforation, Hepatobiliary scan, Infants, Tc<sup>99m</sup>-DISIDA.

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## Introduction

Spontaneous perforation of the common bile duct in infancy is a rare disorder, but it is second to biliary atresia as a cause of surgical jaundice in the infant. First described in 1932, fewer than 150 cases have been reported to date.<sup>(1-3)</sup> The etiology is unknown though distal obstruction and congenital mural weakness in the bile duct wall have been postulated. Trauma, ischemia, distal biliary obstruction, and pancreatic reflux appear also to be causative etiologies.<sup>(1)</sup> We present a case of Spontaneous common bile duct perforation (SBP) in a 5-month-old male baby which was clearly demonstrated by Tc<sup>99m</sup> Disofenin (Tc<sup>99m</sup>- DISIDA) hepatobiliary scan.

## Case Report

We report a five-month-old male infant who

was delivered by normal vaginal delivery at 29 weeks of gestation. At birth he was admitted to the neonatal intensive care unit with some respiratory complications due to immaturity. He recovered completely and was discharged in good general condition. He did well and was thriving till he presented at 5 months of age with yellowish discoloration of the skin and sclera, clay colored stool and severe distention of the abdomen, with deterioration in feeding and weight. There was no history of trauma and the infant had been in good health before this.

On physical examination, he was afebrile but distressed. The abdomen was distended with shifting dullness and 43cm girth. There was bilateral huge hydrocele.

Total serum bilirubin was 17 mg/dl, direct bilirubin 7 mg/dl, alkaline phosphatase (ALP) 1531 units/L, ALT 54 units/L, total protein 56

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g/L, albumin 33 g/L, cholesterol, and 0.7mmol/L. The prothrombin time (PT) was 15s and the partial thromboplastin time (PTT) was 44s and the INR 1.2. Urine was dark color with normal analysis, and the stool was free of bilirubin. His Hemoglobin was 9.1g/dl, WBC 7620/uL and platelets 700,000 /uL. A diagnostic peritoneal tap revealed greenish fluid. Analysis of the ascitic fluid revealed WBC at 60 with 79% polymorphonuclear cells, 18% lymphocytes, and cholesterol at 7.4 mM. Gram stain and cultures for the blood and ascetic fluid were negative. Finally Hepatitis B surface Antigen (HBs Ag) and Hepatitis C Virus Antibodies (HBV Ab's) were negative.

The infant's chest X-ray was normal. An ultrasound of the abdomen showed a large amount of free fluid with a slightly small echogenic liver. Neither the gallbladder nor the biliary tract were seen, both kidneys and spleen appeared normal.

Hepatobiliary radionuclide imaging using an age-weight-adjusted dose of Tc99m-Disofenin was done. Dynamic images with 1 second frames for 60 seconds followed by 1 minute frames for 60 minutes were obtained. Two, 4 and 24 hour delayed images were acquired. There was good prompt hepatic perfusion and uptake of the radiopharmaceutical. In sequential images and delayed images the activity was noted to accumulate in the paracolic gutters bilaterally. On delayed images liver uptake was reduced and there was no activity noted in the bowel, with prominent activity in the peritoneal cavity indicating activity in the free ascetic fluid accumulating in the paracolic gutters (Fig. 1). The impression of biliary leak due to extrahepatic biliary perforation was suggested. This was further proved by positive bilirubin tested in ascetic fluid sample.

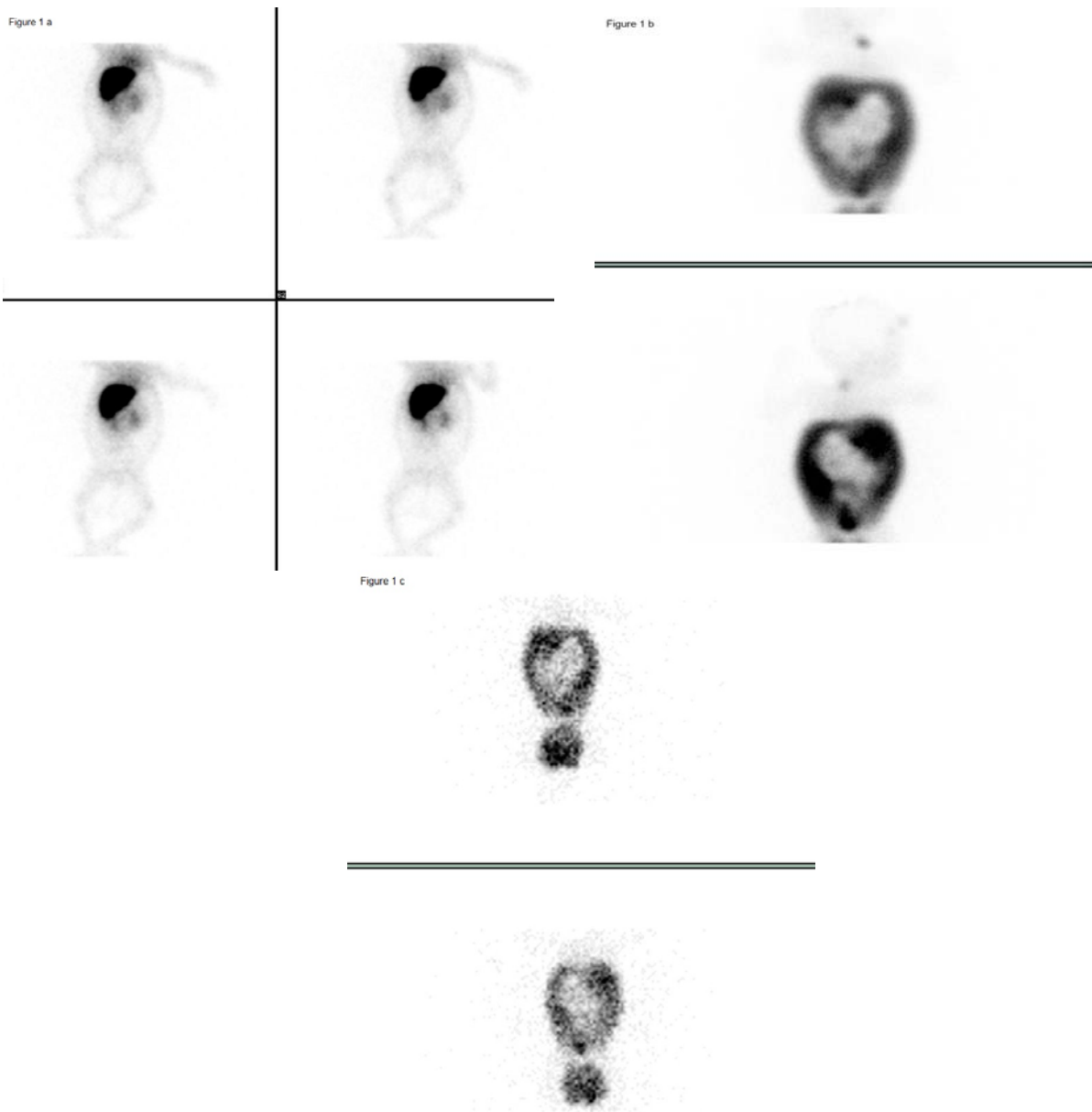
The patient was sent to laparotomy. About one liter of ascetic fluid was drained from the abdominal cavity. A tear in the proximal part of the common bile duct was identified and repaired. A drain was inserted. Repair of small umbilical hernia and bilateral inguinal hernias was done. Liver biopsy revealed macrovesicular fat deposition with normal architecture consistent with moderate fatty infiltration of the liver. Peritoneal biopsy was consistent with biliary peritonitis.

The recovery of the patient was smooth during the first few days post operation. The abdomen was soft and lax, his colour was clearing. One week post operation, unfortunately, the child started to deteriorate. He was febrile, distressed, fatigued, blood culture was positive for Candidal infection. Prompt aggressive antibiotic and antifungal treatment was started, but he rapidly deteriorated and cardiopulmonary arrest caused death of the infant.

## Discussion

Since the first report describing spontaneous rupture of bile duct in an infant, a number of cases have been reported.<sup>(1-4)</sup> The majority of cases of this rare cause of biliary ascites occur in the first 5 months of life;<sup>(1,5)</sup> few cases are reported in older infants, and it is extremely rare in adults.<sup>(6-8)</sup> In adults the usual cause is secondary to gallstones and almost always extrahepatic.<sup>(6,8)</sup> There are also reported cases of spontaneous biliary duct perforation in pregnant females.<sup>(7,9)</sup> The presentation may be as an uncommon acute form or a classic subacute type, and might rarely present with acute abdomen and shock.<sup>(10)</sup> The presenting symptoms, clinical features, and early course of the disease in our patient match those reported earlier.<sup>(1,4,11)</sup> Typically, an infant will develop mild jaundice and anorexia after a period of good health. These symptoms are usually followed by progression to weight loss and subsequent failure-to-thrive. During the next 3-6 weeks, intermittent vomiting develops and the stool becomes pale.<sup>(10)</sup> Progressive abdominal distention associated with umbilical and inguinal hernias follow.<sup>(4)</sup>

Tc<sup>99m</sup>-DISIDA cholescintigraphy has been found to be sensitive in demonstrating spontaneous biliary perforation.<sup>(4,7,11)</sup> Disofenin has about 88% hepatic excretion. This radiopharmaceutical enters the anion exchange pathway of bilirubin and share the same hepatic uptake and excretion pathway. It is taken up by hepatocytes and not conjugated prior to its excretion. In our patient we report a distinct technetium disofenin imaging pattern of radioactivity leakage into the mesenteric folds in the abdomen, and collecting in the paracolic gutter areas, caused by the perforated extrahepatic bile duct.



**Fig. 1:** Sequential Tc disofenin images and delayed 2, 24 hours scans (DISIDA scan):

- a) Early flow images show prompt good hepatic uptake of tracer with no evidence of any excretion in the bowel.
- b) Delayed 2 hours images show progressive leakage of radioactivity along the mesenteric root and mesocolon boundaries into the para-colic gutters.
- c) Filling of gutters filled by ascetic fluid with radioactivity is noticeable at the periphery of the peritoneal cavity even in 24 hour images with reduced uptake in the liver.

It has been postulated that the pathogenesis of spontaneous common duct perforation in infancy could be due to a localized embryogenic malformation, congenital thinning (mural weakness) of the anterior bile duct wall at the junction of the bile and cystic ducts,<sup>(1)</sup> or ischemia, or gall stones.<sup>(6, 9)</sup> No common etiological factors have been identified for the perforations, but they are almost always located near the union of the cystic duct to the common

bile duct, suggesting that this junction may be particularly vulnerable to injury or errors in development.<sup>(11)</sup>

The Hepatobiliary scan was diagnostic in our patient. The lack of drainage of radioactivity into the bowel and the presence of leakage from the common bile duct into the abdomen indicated perforation. Intraoperative cholecystogram could be used when the size of the perforation is too small to be identified visually during the

operation, which was not the case in our patient.<sup>(12)</sup>

The usual course after surgery and repair of the perforation is recovery and resolution of the jaundice, normal color of stool and urine and improvement in the condition of the infant with regard to feeding and thriving. Later, a repeat hepatobiliary scan shows adequate liver uptake and biliary drainage to the bowel.<sup>(1-4,7,13)</sup> Unfortunately, the course in our reported case was complicated, though transient improvement in the condition occurred, by Candidal septicemia documented after a week of operation. Eventually septic shock and death occurred. The long-term prognosis is usually excellent when the clinicians are aware of this rare surgically corrected entity, and the problems encountered are usually due to complications of infection in a poorly thriving infant.

## Conclusion

Spontaneous perforation of the common bile duct in infancy is a rare disorder, yet second to biliary atresia as a cause of surgical jaundice in infancy. Many lab and diagnostic tools are useful in the diagnosis of this entity. We emphasize the importance of Tc<sup>99m</sup> Disofenin (Tc<sup>99m</sup>- DISIDA) hepatobiliary scan in establishing the diagnosis of this rare, yet surgically correctable disease.

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