

# Portal cholangiopathy: case report\*

*Colangiopatía portal: relato de caso*

**Maria Cecília Almeida Maia<sup>1</sup>, Aline Pimentel Amaro<sup>1</sup>, Edmundo Clarindo Oliveira<sup>2</sup>, José Renan da Cunha Melo<sup>3</sup>, Marcelo Dias Sanches<sup>3</sup>, Rogério Augusto Pinto da Silva<sup>1</sup>**

Maia MCA, Amaro AP, Oliveira EC, Melo JRC, Sanches MD, Silva RAP. Portal cholangiopathy: case report. Radiol Bras. 2014 Jan/Fev;47(1):51-53.

**Abstract** The present report describes the case of a child that after blunt abdominal trauma presented with portal thrombosis followed by progressive splenomegaly and jaundice. Ultrasonography and percutaneous cholangiography revealed biliary dilatation secondary to choledochal stenosis caused by dilated peribiliary veins, characterizing a case of portal biliopathy. The present case report is aimed at presenting an uncommon cause of this condition.

**Keywords:** Portal cholangiopathy; Portal biliopathy; Portal hypertension; Portal vein cavernous transformation.

**Resumo** Relata-se o caso de uma criança que após trauma abdominal fechado apresentou trombose portal, seguida por esplenomegalia progressiva e icterícia. Os achados da ultrassonografia e da colangiografia percutânea mostraram dilatação de vias biliares secundária à constrição do colédoco por veias pericoledocianas dilatadas, configurando caso de colangiopatía portal. O objetivo deste relato é a apresentação de causa incomum desta condição.

**Unitermos:** Colangiopatía portal; Biliopatía portal; Hipertensão portal; Transformação cavernomatosa da veia porta.

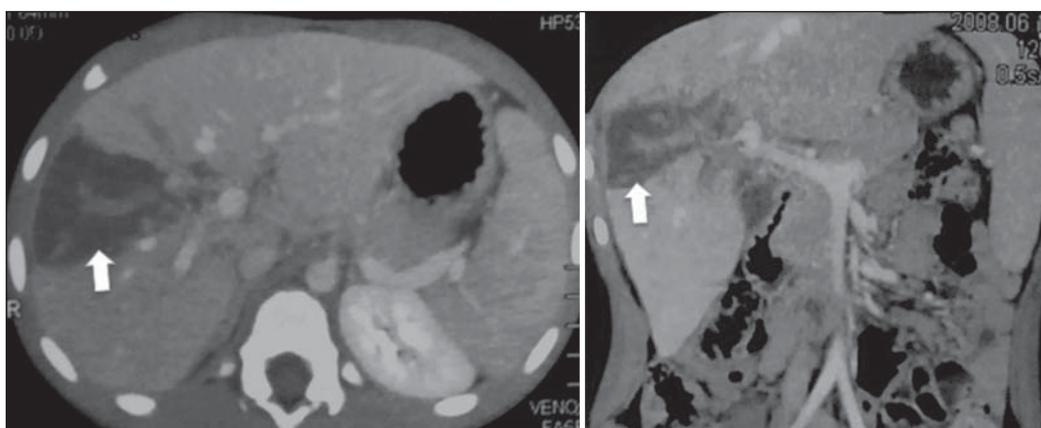
## INTRODUCTION

Portal cholangiopathy is defined as obstruction of the biliary tract in association with engorgement of collateral veins in the hepatic hilum<sup>(1)</sup>. In general, the patients are asymptomatic, but jaundice, cholangitis and biliary lithiasis may be present in 5% to 38% of patients<sup>(1-5)</sup>. In the present report, the authors describe the case of a child with portal thrombosis secondary to blunt abdominal trauma, progressing with portal hypertension and obstructive jaundice caused by portal biliopathy.

## CASE REPORT

In May 2008, at the age of six years and seven months, a male child suffered an accident and had fracture of left lower limb and blunt abdominal trauma, with injury to the right liver lobe (Figure 1), which were conservatively treated.

In March 2009, the patient presented with splenomegaly. Abdominal ultrasonography revealed total thrombosis of the right portal vein branch and stenosis between the intrahepatic portal vein and the left branch (Figure 2). An unsuccessful attempt was made for dilatation of the stenosis which



**Figure 1.** Computed tomography (axial and coronal reconstruction) performed at hospital admission, showing hepatic trauma affecting the right portal branch and the hepatic hilum (arrows). The portal vein and the left portal branch were not injured at that time.

\* Study developed at Clínica CEU Diagnósticos, Belo Horizonte, MG, Brazil.

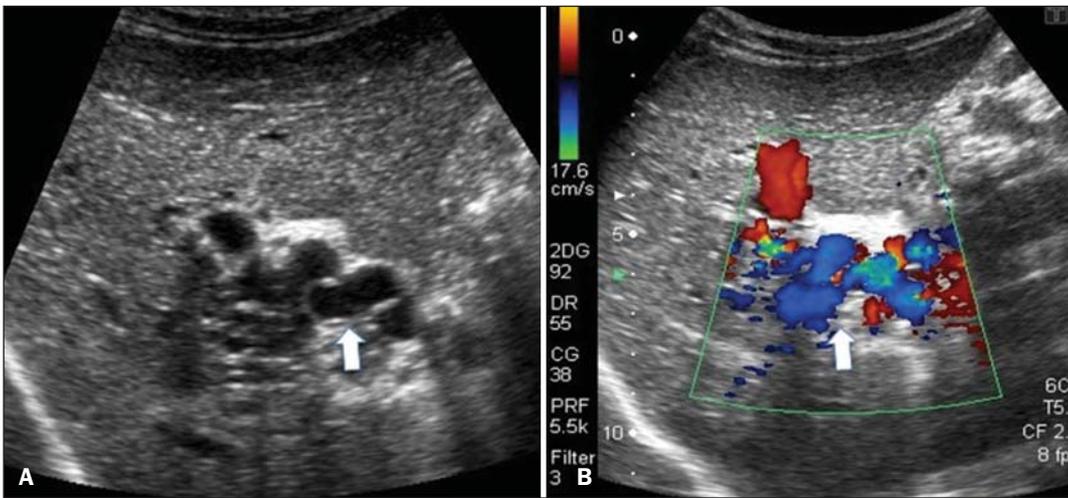
1. MDs, Radiologists at Clínica CEU Diagnósticos, Belo Horizonte, MG, Brazil.

2. MD, Hemodynamicist at Clínica CEU Diagnósticos, Belo Horizonte, MG, Brazil.

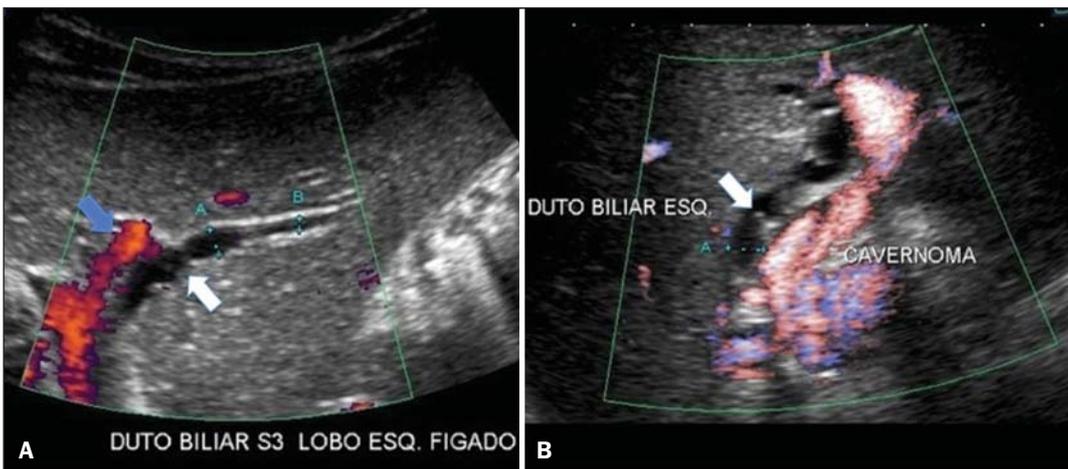
3. PhDs, Professors, Department of Surgery at School of Medicine, Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, MG, Brazil.

Mailing Address: Dra. Maria Cecília Almeida Maia. Rua Oscar Trompowsky, 721/406, Gutierrez. Belo Horizonte, MG, Brazil, 30441-055. E-mail: mariaceciliaalmeida@yahoo.com.br.

Received November 12, 2012. Accepted after revision May 27, 2013.



**Figure 2.** Ultrasonography (A) and color Doppler mapping (B) identifying multiple tortuous periportal veins in the hepatic hilum (arrows) corresponding to cavernous transformation of the portal vein.



**Figure 3.** Dilatation of the biliary tract involved by prominent venous collaterals. Dilated left bile duct (white arrows and electronic pakymeters). Distal portion of the left branch of the patent portal vein (blue arrow). Cavernoma refers to tortuous periportal veins in the hepatic hilum.

progressed to total occlusion of the portal vein bifurcation and progressive splenomegaly as well as venous engorgement of collaterals surrounding the portal vein from the head of pancreas to the hepatic hilum.

In December 2010, the patient presented with jaundice, and in January 2011, ultrasonography demonstrated intrahepatic biliary dilatation in association with presence of calibrous periportal veins (Figure 3).

Echo-guided percutaneous puncture of biliary duct and subsequent cholangiography were performed, demonstrating reduction of the choledochal caliber. Later, Doppler US revealed the engorged vein twirled around the biliary drain into the choledochus (Figure 4).

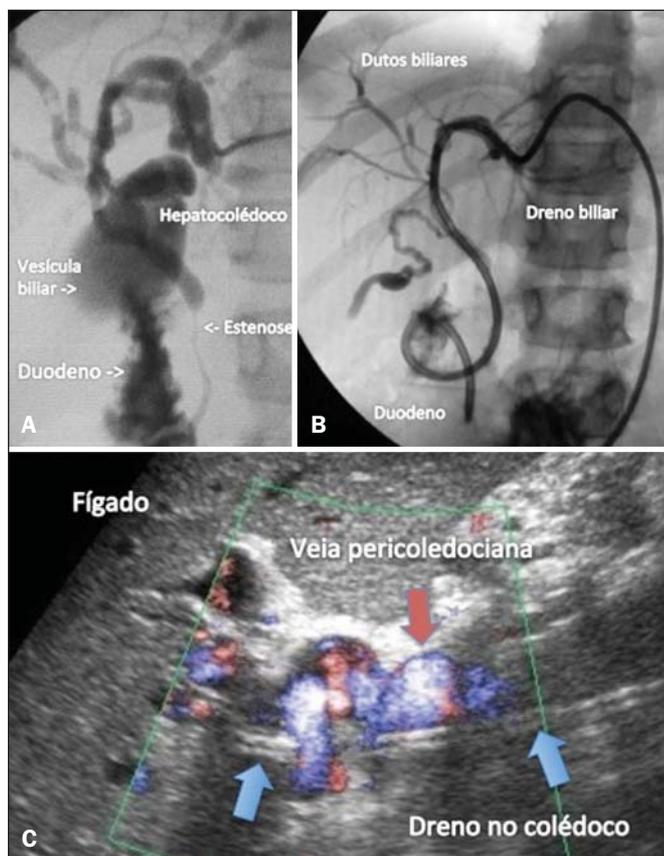
New attempts to dilate the stenosis on March 19, May 21, 2011, and March 10, 2012, with persistence of the extrinsic compression. On July 14, 2012, percutaneous puncture of the spleen for splenic vein catheterization and cannulation of the proximal segment of the portal vein and of the pericholedochal collateral were done, but the attempt was unsuccessful.

Angiography demonstrated obstruction of the portal vein after its origin, in association with significant dilatation of pericholedochal veins up to the hepatic hilum, followed by

enhancement of many small and thin veins similar to a cloud, with delayed filling of the left portal branch. The anatomy and pressure in the hepatic veins were normal. Several guide wires were utilized in the attempt to recover the lumen of the portal vein or to get the thin veins of the hepatic hilum dilated, but such an attempt was also unsuccessful. Currently, the child remains with an internal-external type biliary drain that is periodically replaced, with assessment by means of cholangiography.

## DISCUSSION

Portal cholangiopathy results from alterations of the biliary tract in the cavernous transformation of the portal vein<sup>(6)</sup>, in association with engorgement of periportal veins around the pancreas, choledochus and hepatogastric ligament<sup>(5)</sup>. As the veins of the epicholedochal plexus (plexus of Saint) and paracholedochal plexus (plexus of Petren) respectively located around and on the wall of the hepatocholedochus are engorged, one may observe its compression also associated with the inflammatory ischemic process which causes areas of fibrosis with consequential biliary tract stenosis which resolve after the management of the portal hypertension<sup>(7)</sup>.



**Figure 4. A:** Percutaneous cholangiography demonstrating dilatation of the biliary tract up to the common bile duct, with abrupt decrease in the caliber at the level of the middle third of the choledochus. **B:** Internal-external-type drain percutaneously inserted into the biliary tract. **C:** Doppler ultrasonography demonstrating pericoledochal vein around the drain (arrow) in the hepatocholedochus.

In spite of such a condition being frequently observed at imaging methods, the patients, particularly children, are generally asymptomatic, eventually presenting increased levels of hepatic enzymes.

In the present case report, the authors describe the set of symptoms of a child with portal biliopathy secondary to portal vein thrombosis, consequential to blunt abdominal trauma, developing with portal hypertension and obstructive jaundice refractory to treatment.

The clinical cholangiopathy manifestations can be divided into two groups, as follows: a) those resulting from cholestasis (jaundice and itch); b) those secondary to intrahepatic biliary lithiasis, corresponding to pain in the right hypochondrium, jaundice and fever. Symptoms are most commonly observed in the elderly, particularly in the presence of gallbladder calculi.

Morphological alterations most frequently observed at cholangiography include biliary tract dilatation associated with areas of stenosis and angulations, as well as failure in duct filling due to compression by dilated parietal veins projecting towards the lumen<sup>(6)</sup>.

Differential diagnosis should include hepatic hilum neoplasia, such as cholangiocarcinoma or metastatic neoplasms, as well as other causes of intrahepatic bile ducts involvement, such as sclerosing cholangitis.

The diagnosis is made by imaging methods, particularly color Doppler that is capable of detecting portal thrombosis and bile ducts dilatation. In cases of portal vein fibrosis caused by chronic portal thrombosis, it usually cannot be seen at B-mode, while the collaterals are visualized as serpiginous structures in the hepatic hilum, with venous flow at Doppler.

Magnetic resonance imaging also allows the diagnosis of cavernous transformation of the portal vein and biliary dilatation, particularly by means of MR cholangiography.

Cholangiography (either retrograde or percutaneous) may be done in cases where the treatment is indicated for dilatation and drainage of the biliary ducts as well as for decompression of portal system. If available, echoendoscopy represents an alternative method, allowing the study of the biliary tract and, depending on the equipment, it allows the utilization of the Doppler technique.

As regards treatment, bile duct dilatation is indicated for symptomatic patients and with recurrent cholangitis. Portal decompression should generally be performed before bile ducts dilatation<sup>(6)</sup>.

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