

Images in Hepatology

Caroli's Disease and Choledochal Cyst

Jorge Hérnandez-Ortiz MD, 1 Roberto Corona MD, 1 Nahum Méndez-Sánchez MD PhD1

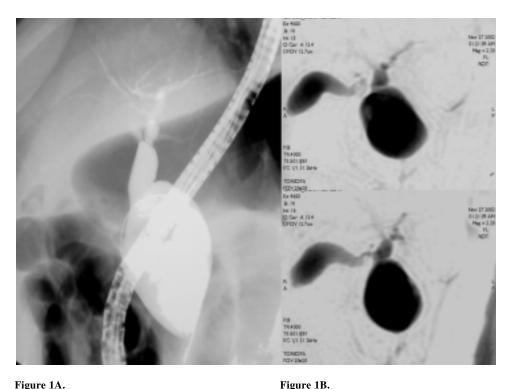


Figure 1A.

A 33-year old female was admitted to the hospital presented with right hypochondrium abdominal pain and jaundice. Abdominal ultrasonography showed multiple saccular dilatations of the intrahepatic biliary tract. Endoscopic Retrograde Cholangiopancreatography was performed (Figure 1A) showing nonobstructive saccular or fusiform dilatation of the intrahepatic bile ducts and choledochal cyst. Also it was performed a MR cholangiography (Figure 1B) which showed the same abnormalities described above.

In 1958 Jacques Caroli described communicating cavernous ectasia of the biliary tree as an uncommon cause of chronic, often life-threatening hepatobiliary disease. The disease now most often referred to as Caroli's disease is a

rare condition characterized by nonobstructive saccular or fusiform dilatation of the intrahepatic bile ducts. In the socalled pure form, dilatation is classically segmental and saccular and is associated with stone formation and recurrent bacterial cholangitis. In the form associated with congenital hepatic fibrosis, bile duct dilatation usually is less prominent; portal hypertension and eventual liver failure typically develop as a result of the hepatic fibrosis. Caroli's disease usually is manifested in childhood and is thought to be congenital and probably inherited. Associated conditions include renal cystic disease, choledochal cysts, and cholangiocarcinoma. This pictorial essay illustrates the broad spectrum of imaging findings in Caroli's disease.

Address for correspondence: Jorge Hernández-Ortiz MD Medica Sur Clinic & Foundation Puente de Piedra 150, Col. Toriello Guerra, Mexico City, Mexico

References

- Miller WJ, Sechtin AG, Campbell WL, Pieters PC. Imaging findings in Caroli's disease. AJR 1995;165:333-7.
- Krause D, Cercueil JP, Dranssart M, Cognet F, Piard F, Hillon P.MRI for evaluating congenital bile duct abnormalities. J Comput Assist Tomogr 2002;26:541-52.

¹ Departments of Radiology, Gastroenterology and Liver Unit.