

ANNALS OF HEPATOLOGY




Volume **1** Number **4** October-December **2002**

Artículo:




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Images in Hepatology

Caroli's Disease and Choledochal Cyst

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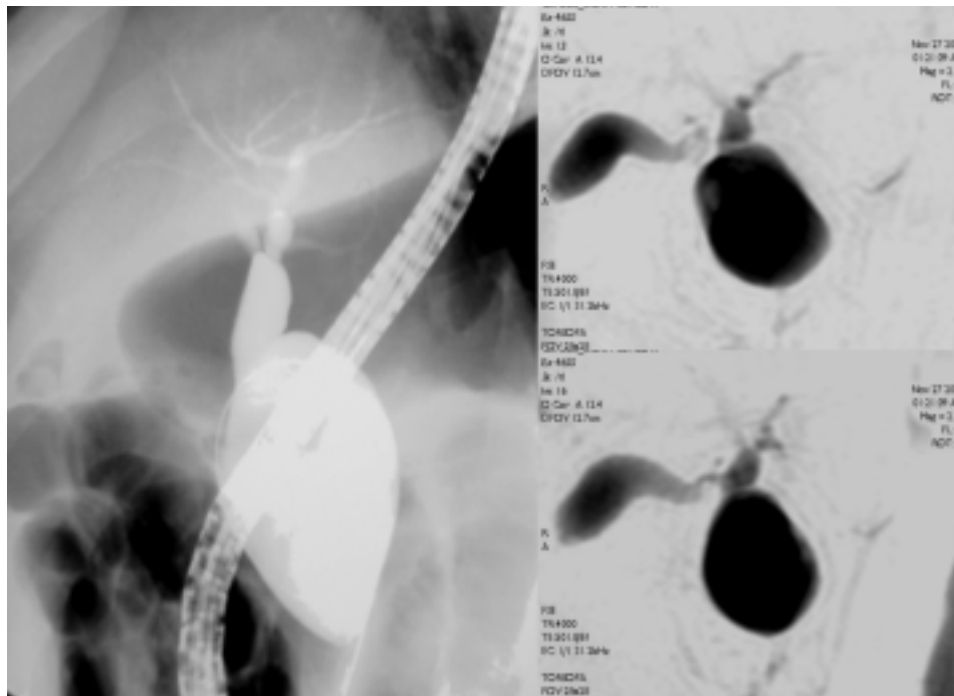


Figure 1A.

Figure 1B.

A 33-year old female was admitted to the hospital presented with right hypochondrium abdominal pain and jaundice. Abdominal ultrasonography showed multiple saccular dilations 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 so-called 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.

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