

## Adolescent Upper GI Bleed: A Rare Cause

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### Abstract

This is a case of an adolescent male presenting with recurrent episodes of well tolerated upper gastrointestinal bleed. There was no history of jaundice during the index episode or in past. There was no history of umbilical vein sepsis at birth/growth retardation/short stature and no significant family history. Patient had hepatosplenomegaly with anemia and pancytopenia. Upper gastrointestinal endoscopy revealed large esophageal varices. Non-cirrhotic portal hypertension is more common cause of adolescent upper gastrointestinal bleed than cirrhotic portal hypertension. USG examination revealed multiple cystic/hypoechoic lesions in both lobes of liver and hepatosplenomegaly. Patient underwent magnetic resonance cholangiopancreatography which revealed an uncommon cause of upper GI bleed.

**Keywords:** Adolescent; Upper GI Bleed; Magnetic Resonance Cholangiopancreatography

### Introduction

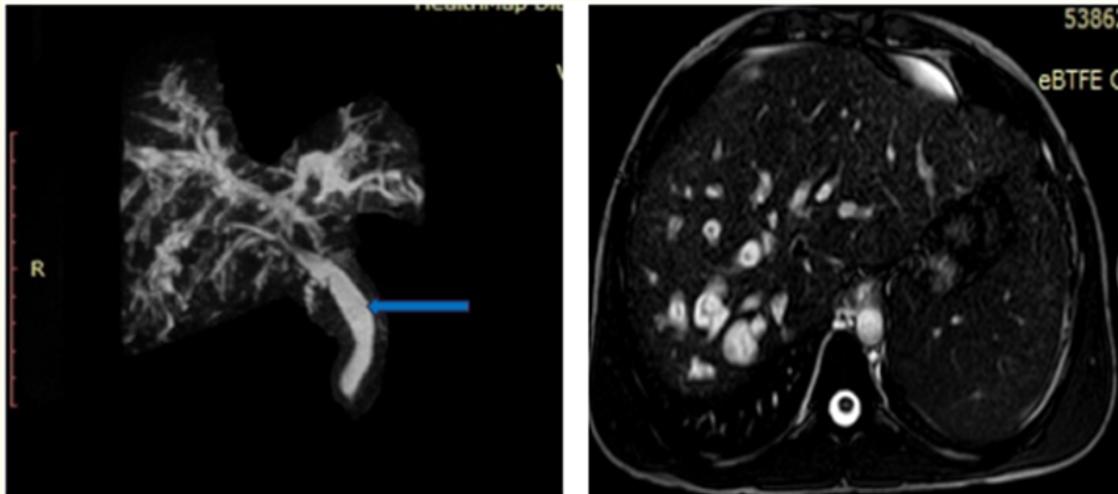
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### Case Report

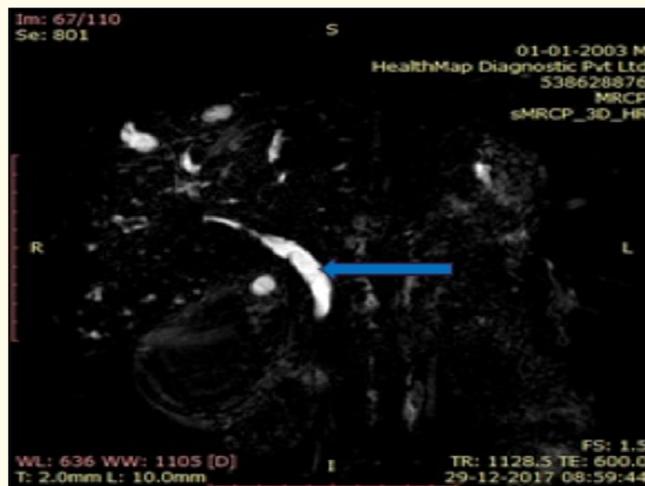
A 15-year male presented with painless, profuse, well tolerated episodes of upper gastrointestinal bleed since 3 years. He had no history of jaundice, pruritus, fever or abdominal distension. On general physical examination, he had hepatomegaly with liver palpable 6 cm below right costal margin and mild splenomegaly. On investigations he had pancytopenia with Hemoglobin 6.1 gm/dL, total leukocyte count 1300/cumm, Platelet count 75,000/cumm with mildly raised liver enzymes SGOT 59 U/L, SGPT 40 U/L, ALP 102 U/L, Total Bilirubin 0.3 mg/dL, prolonged prothrombin time 20 sec with INR 1.47. His upper gastrointestinal endoscopy showed high grade varices for which endoscopic band ligation was performed. USG abdomen showed hepatosplenomegaly with hypoechoic and cystic lesions in both lobes of liver. Etiological work up revealed negative viral markers, normal ceruloplasmin, normal 24-hour urinary copper, no KF Ring and negative autoimmune markers with normal total IgG levels.

Magnetic resonance cholangiopancreatography (MRCP) showed focal tubular and cystic dilatations of intrahepatic biliary radicles (IHBR) in both lobes of liver. A central dot like hypointensity was seen in many of these dilated ducts (Figure 1). No significant dilatation of extrahepatic common bile duct (CBD) is seen with smooth tapering of distal CBD (Figure 2). The maximum intensity projected image

also reflected the same findings showing cystic and tubular dilated IHBR and no significant dilatation of CBD with smooth tapering of CBD. No filling defect could be seen in CBD. The gall bladder is not seen distended. There is also splenomegaly. These findings were typical for caroli disease which is a type V choledochal cyst.



**Figure 1:** Magnetic resonance cholangiopancreatography (MRCP) showed focal tubular and cystic dilations of intrahepatic biliary radicles (IHBR) in both lobes of liver with a central dot like hypointensity in many of these dilated ducts.



**Figure 2:** No significant dilatation of extrahepatic common bile duct (CBD) with smooth tapering of distal CBD.

## Discussion

The hepatic fibrocystic diseases are congenital disorders due to segmental, saccular dilatation of intra or extrahepatic biliary system. Caroli disease is a developmental anomaly of ductal plate formation during early intrauterine gestation although can occur in late gestational period also. Caroli disease involves saccular and segmental dilatation of intrahepatic biliary radicles [1,2]. These radicles are lined by biliary epithelium which during episodes of cholangitis, is infiltrated by acute and chronic inflammatory infiltrate. There may be formation of small stones leading to microlithiasis which further increases the risk of secondary biliary cirrhosis. There can be varying presence of portal fibrosis. If caroli disease is associated with congenital hepatic fibrosis (CHF), it is called caroli syndrome [3]. The presence of CHF signifies onset of early portal hypertension in these patients. The expansion of portal fibrosis with hypoplastic portal vein branches is not associated with regeneration in surrounding liver parenchyma, differentiating congenital hepatic fibrosis from cirrhosis of liver. Patient may present with esophageal varices with or without ascites. Caroli disease increases the risk of future development of cholangiocarcinoma. Our patient presented with recurrent episodes of UGI bleed and pancytopenia suggesting hypersplenism due to portal hypertension. A diagnosis of caroli disease with congenital hepatic fibrosis was made indicating Caroli syndrome. However, index patient never had any episode of jaundice/cholangitis.

In management of caroli disease, episodes of recurrent cholangitis should be treated by antibiotics. Endoscopic retrograde cholangiopancreatography (ERCP) is used to extract larger bile duct stones where intrahepatic stones are difficult to manage. Segmental hepatectomy is important treatment option in recurrent cholangitis due to intrahepatic stones. Esophageal varices are managed endoscopically with band ligation. Refractory cases may be dealt with shunt surgery. Overall, if symptoms of recurrent cholangitis are refractory or there is development of secondary biliary cirrhosis, liver transplantation is the only final modality left for these patients [4].

Caroli syndrome is an uncommon cause of portal hypertension in adolescence which require early suspicion and diagnosis so that timely treatment can improve overall outcome of its complications like recurrent cholangitis and variceal bleeding. A significant proportion of patients of caroli disease have autosomal recessive disorder, and mostly this is associated with autosomal recessive polycystic kidney disease [5]. Caroli disease with renal involvement manifests either at birth or early in childhood. In late adolescence and adult life the presentation of caroli disease is usually without renal manifestations.

## Conclusion

Caroli syndrome is an uncommon cause of portal hypertension in adolescence which require early suspicion and diagnosis so that timely treatment can improve overall outcome of its complications like recurrent cholangitis and variceal bleeding. A significant proportion of patients of caroli disease have autosomal recessive disorder, and mostly this is associated with autosomal recessive polycystic kidney disease [5]. Caroli disease with renal involvement manifests either at birth or early in childhood. In late adolescence and adult life the presentation of caroli disease is usually without renal manifestations.

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