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Caroli syndrome with massive polycystic kidney involvement

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Description

A 39-year-old patient with a longstanding history of polycystic kidney disease and cavernous transformation of the portal vein since childhood underwent MR cholangiography. Imaging findings are consistent with Caroli syndrome, demonstrating non-obstructive cystic dilatation of the intrahepatic bile ducts, a choledochal cyst classified as Todani type IVa, hepatic fibrosis with signs of portal hypertension, and associated renal anomalies in the form of polycystic kidneys (Figure 1), Caroli disease is a fibrocystic liver disorder characterized by congenital, non-obstructive cystic dilatation of the intrahepatic bile ducts. When associated with congenital hepatic fibrosis, the condition is termed Caroli syndrome. Caroli syndrome is the more common form and is frequently linked to cystic renal anomalies, most notably autosomal recessive polycystic kidney disease (ARPKD) and Cacchi-Ricci disease, which are present in 60-80% of cases. The condition is typically diagnosed late due to the lack of specific clinical signs. Diagnosis is commonly made during the evaluation of complications, such as intrahepatic lithiasis, cholangitis, hepatic abscesses, portal hypertension, or due to associated conditions, including chronic kidney disease [1]. MRCP is the reference imaging for Caroli disease, offering detailed visualization of the biliary tree.

It detects cystic dilatations, their communication with bile ducts, and the characteristic “central dot sign.” US an CT are less specific; ERCP may serve as a complementary tool [2]. Caroli syndrome treatment includes antibiotics for cholangitis and ursodeoxycholic acid to prevent stones. Surgery or partial hepatectomy is considered in localized forms, while liver transplantation is indicated in diffuse disease with fibrosis or malignancy.

Genetic counseling is essential due to its autosomal recessive inheritance [3].

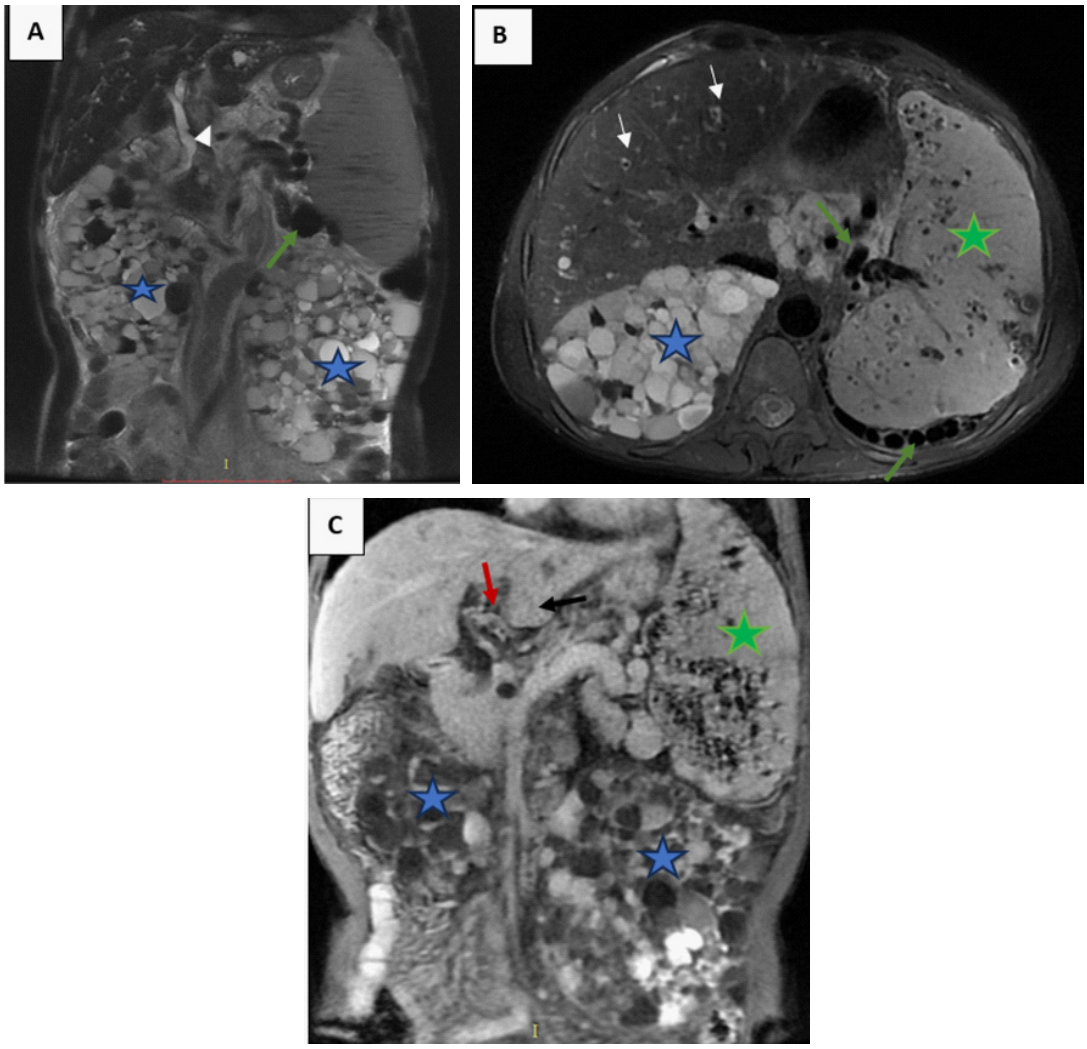

Figure 1: (A–B) Axial and coronal T2-weighted fat-saturated images, and **(C)** coronal post-contrast T1-weighted image showing: A dysmorphic liver with hypertrophy of segment I, (red arrow), and cystic dilatation of several intrahepatic bile ducts containing centrally portal veins (white arrow), forming the characteristic "dot sign," associated with cystic dilatation of the common bile duct (white arrow head), consistent with a Todani type IVa biliary cyst. The portal vein is not visualized and is replaced by multiple periportal venous collaterals, suggestive of cavernous transformation (Red arrow). Both kidneys are markedly enlarged, and are entirely replaced by innumerable cysts, with no visible renal cortex. Some cysts show internal signal heterogeneity compatible with hemorrhagic content (Blue star). The spleen is also enlarged and contains numerous small showing blooming artifacts consistent with Gamna-Gandy bodies (green star). addition, there are multiple direct and indirect portosystemic shunts (green arrow).

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