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Central dot sign

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Caroli disease is a rare, autosomal recessive disorder characterized by communicating cavernous biliary ectasia. Caroli syndrome is a combination of Caroli disease with congenital hepatic fibrosis. In addition, Carolis disease and syndrome can be associated with renal disorders such as medullary sponge kidney, autosomal polycystic kidney disease, and medullary cystic disease [1, 2].

On imaging, Caroli disease demonstrates multifocal, segmental, saccular or fusiform cystic intrahepatic biliary dilatation. These dilated bile ducts can contain calculi or biliary sludge [2]. The Central Dot Sign, representing a portal vein branch protruding into the lumen of a dilated bile duct, can be seen with ultrasound, CT, and MRI (Fig. 1). Presence of the Central Dot sign is highly suggestive of Carolis disease, helping to differentiate it from other causes of intrahepatic biliary dilatation such as primary sclerosing cholangitis and recurrent pyogenic cholangitis. The central fibrovascular bundle enhances after contrast administration [2, 3].

Bile stagnation-related conditions such as stone formation, cholangitis, and liver abscesses can complicate Caroli disease, as can secondary biliary cirrhosis and cholangiocarcinoma [2].

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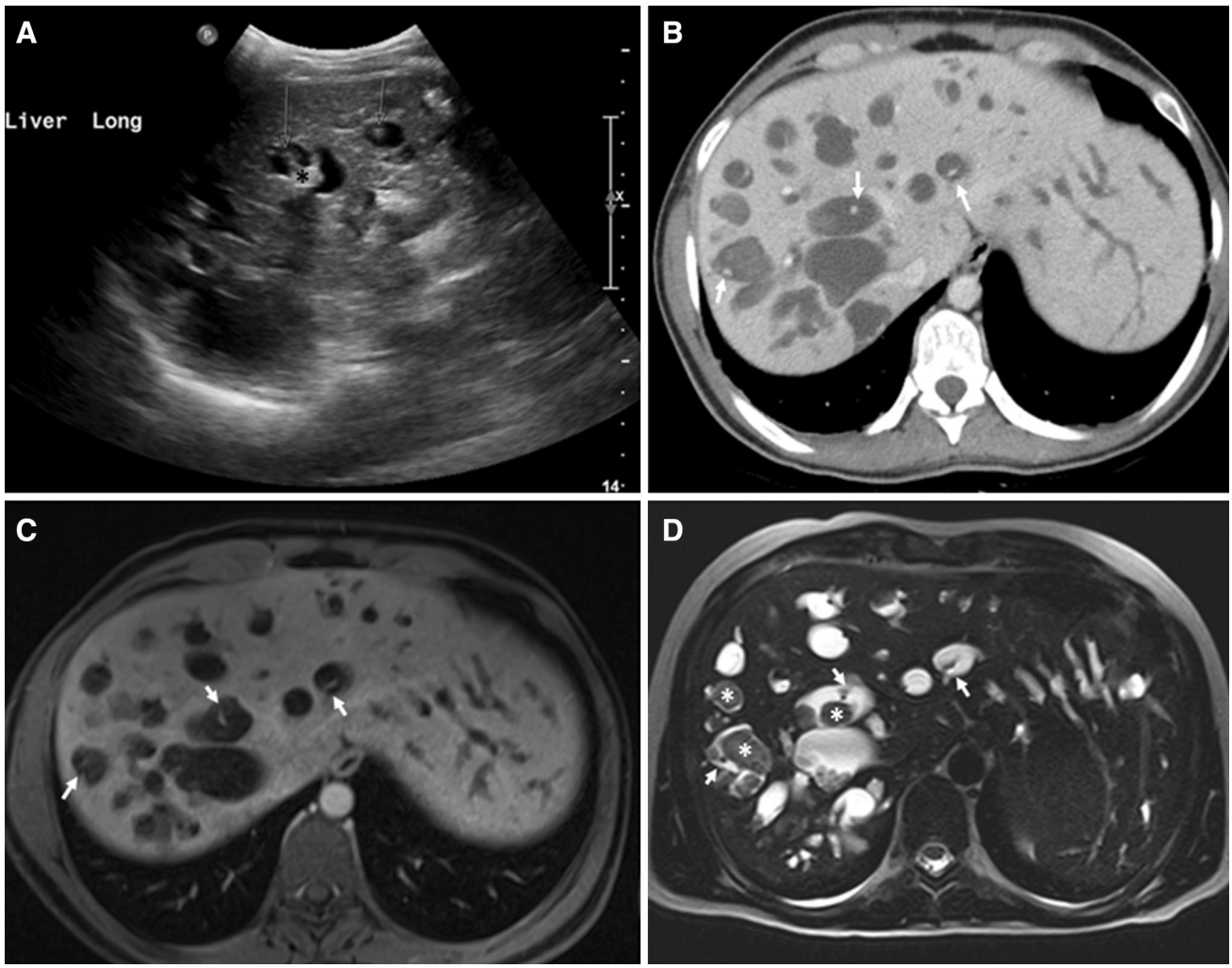

Fig. 1. Longitudinal hepatic sonogram (**A**), axial contrast-enhanced CT (**B**), axial contrast-enhanced T1-weighted MR (**C**), and axial T2-weighted MR images reveal multifocal, segmental, cystic dilatation of the intrahepatic bile ducts with enhancing central dot sign (*arrows*) and multiple calculi (*asterisks*).

Compliance with ethical standards

Conflict of interest All the authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

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