8-year-old boy with history of Caroli syndrome and pancreatitis presenting with epigastric pain and nausea for 1 day

Rohin Thomas, MS3

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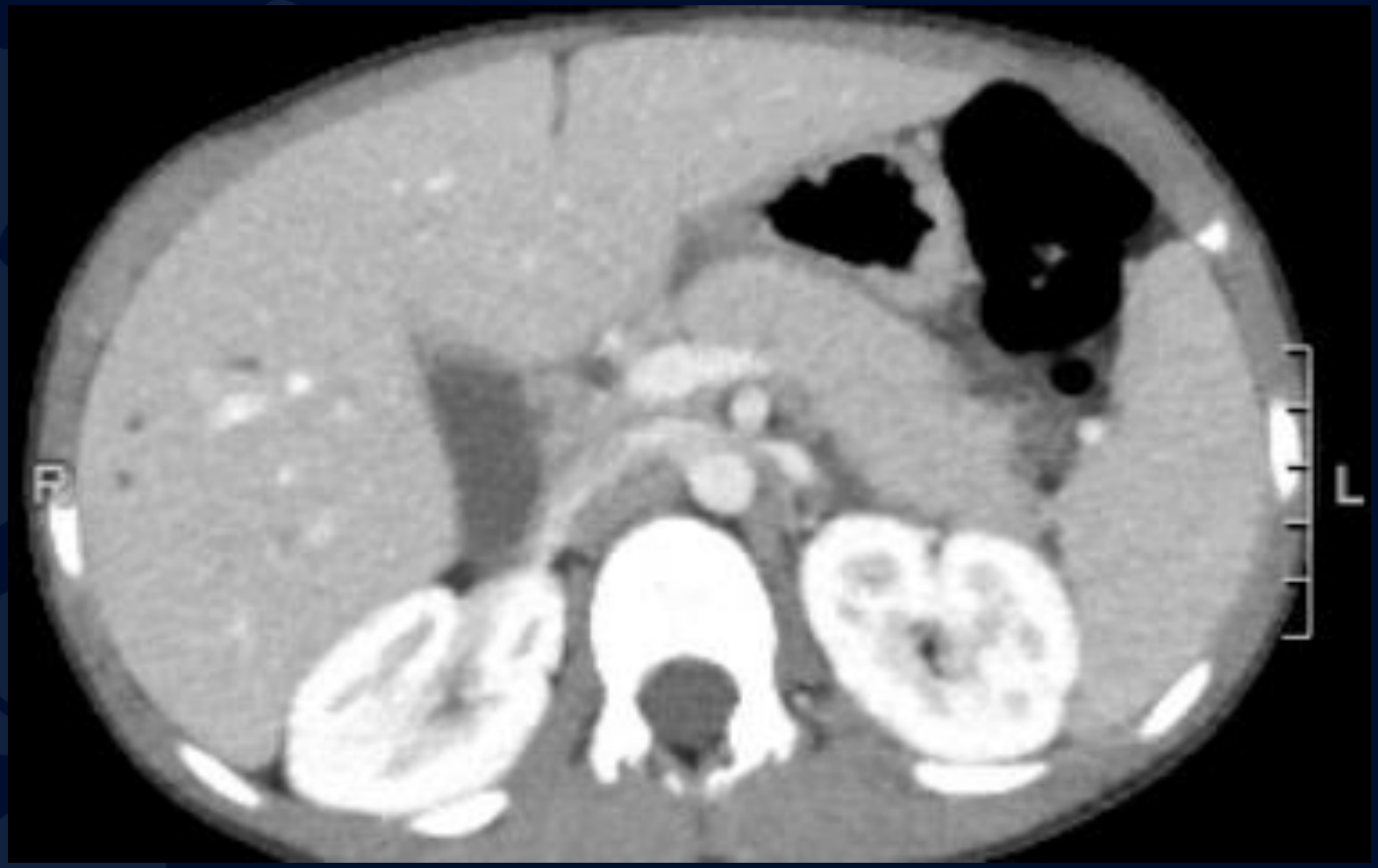
CT IV Contrast



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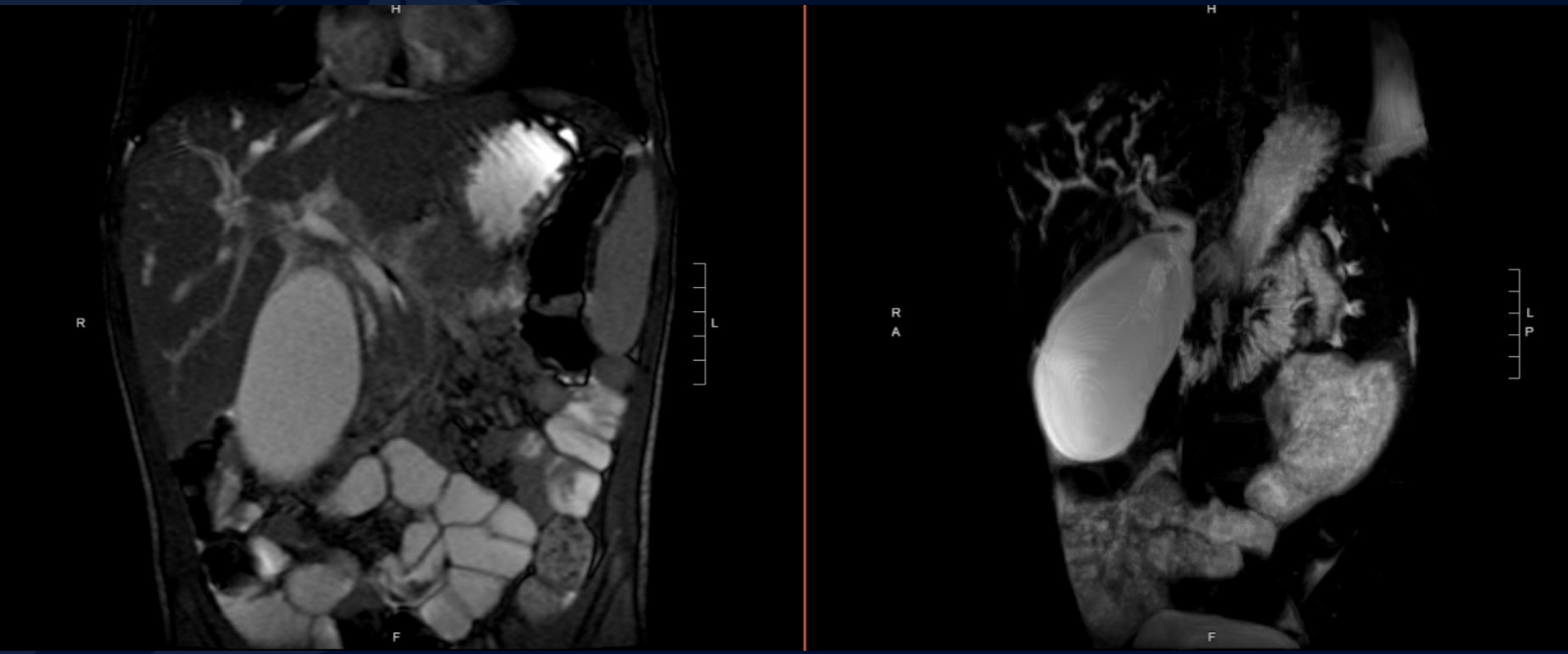
MRCP



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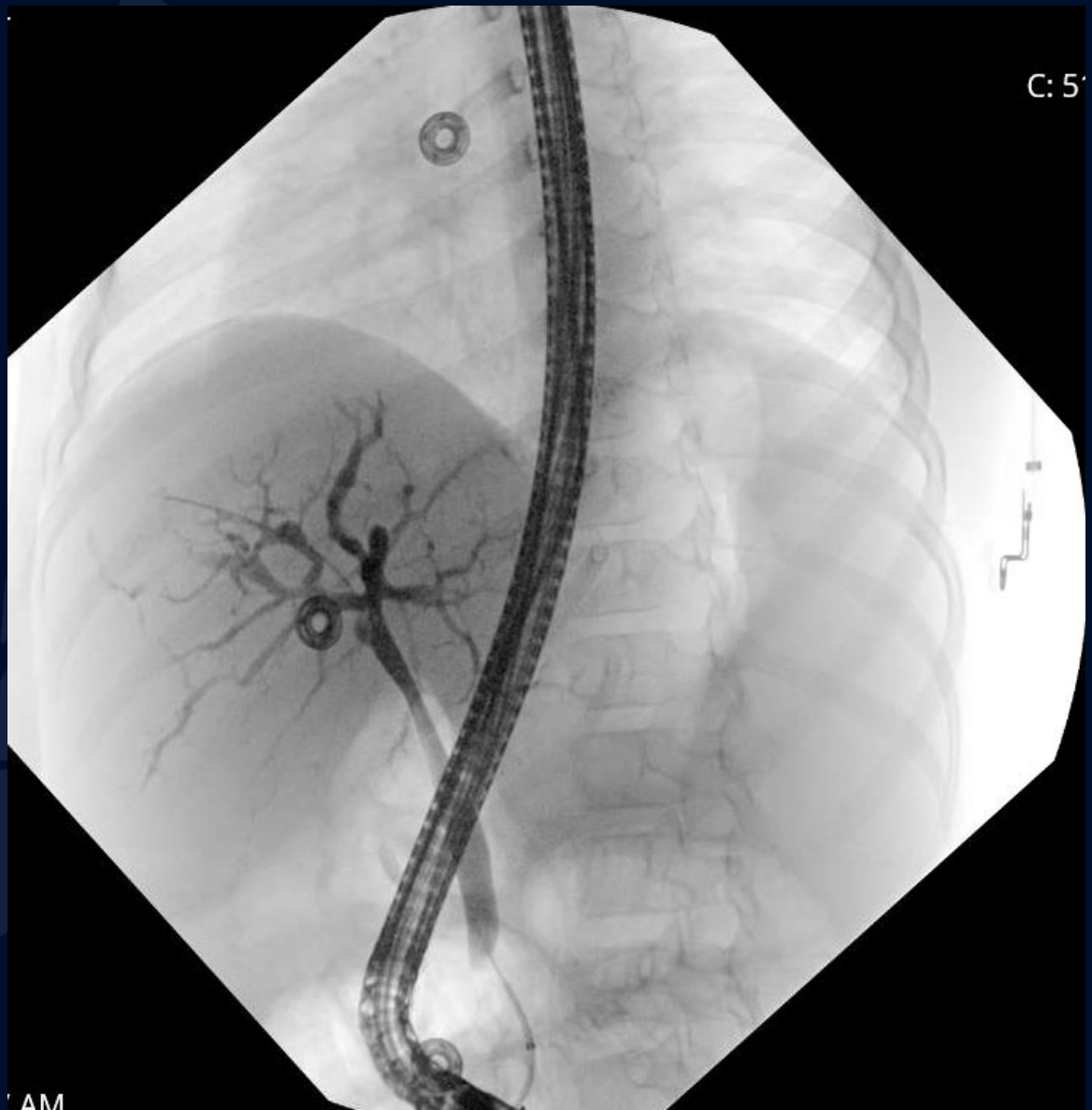
ERCP



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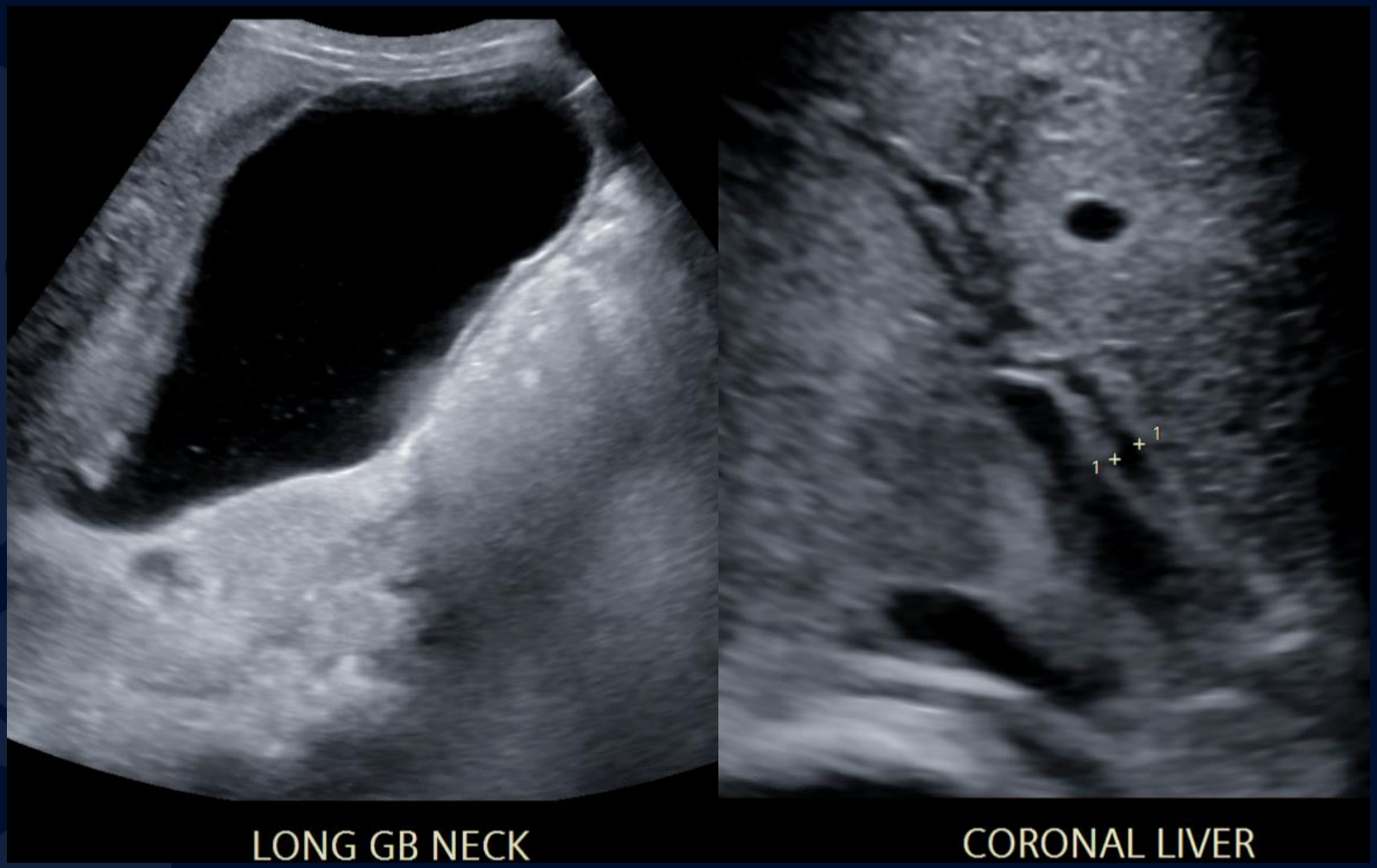
Ultrasound



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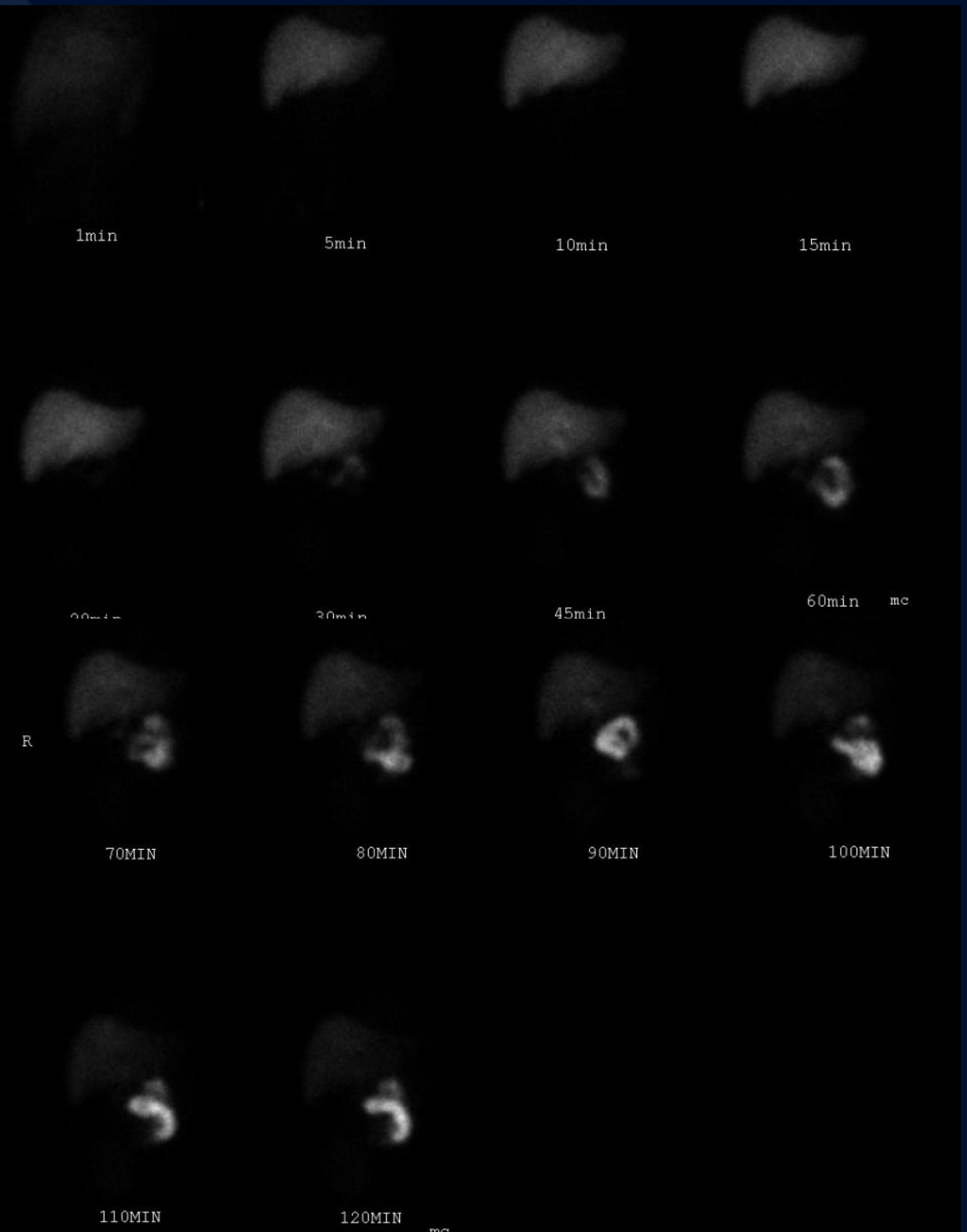

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Acute Biliary Pancreatitis Secondary to Caroli Syndrome

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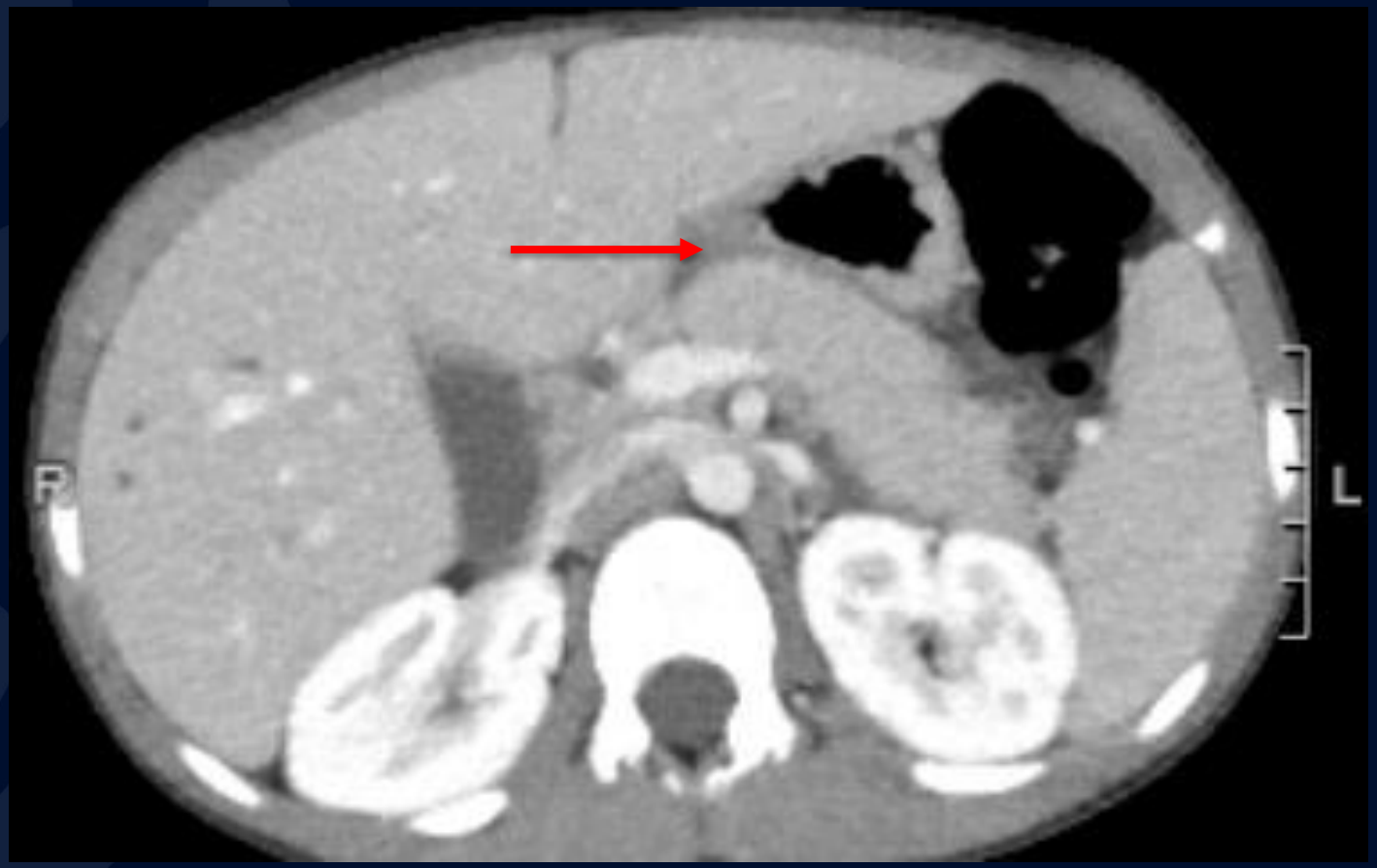
CT IV Contrast



Mild peripancreatic fat stranding

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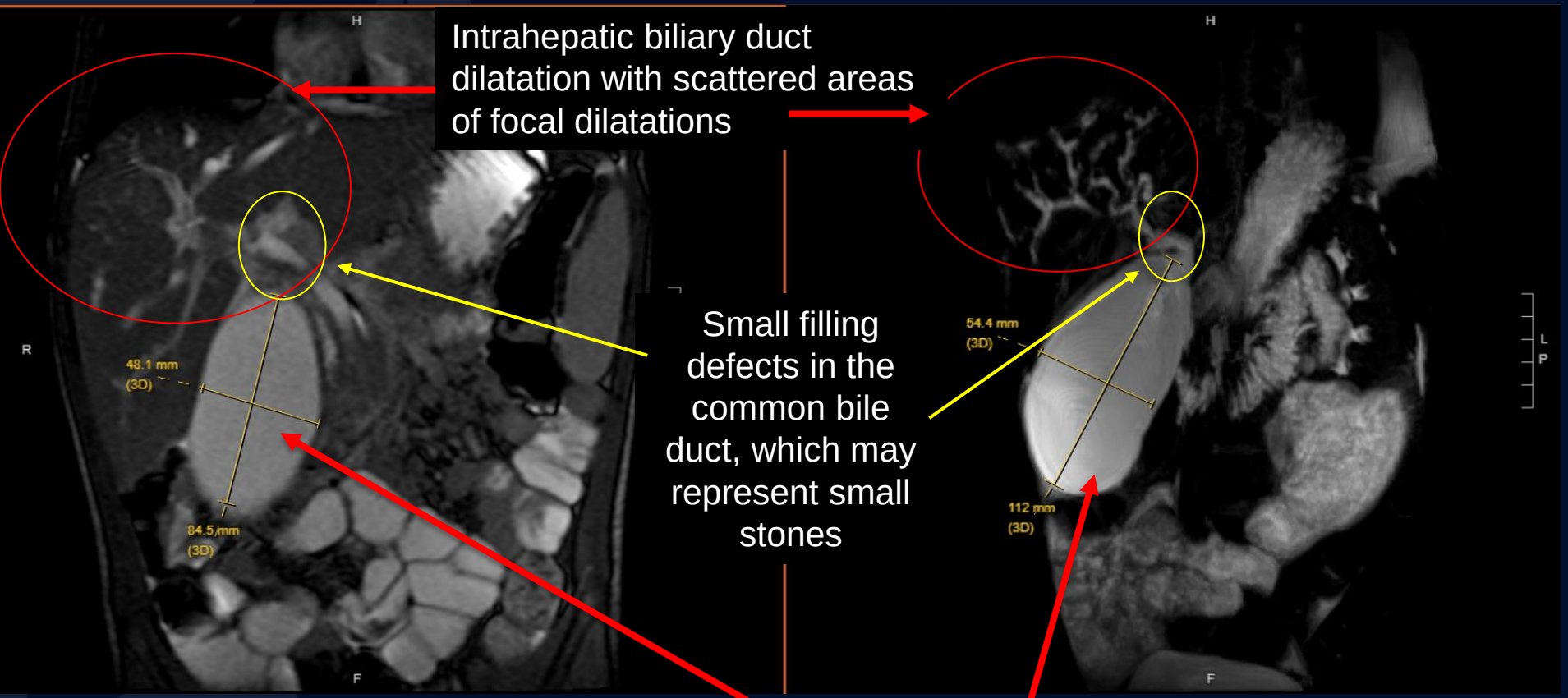

Intrahepatic biliary duct dilatation with scattered areas of focal dilatations

Small filling defects in the common bile duct, which may represent small stones

Markedly distended gallbladder

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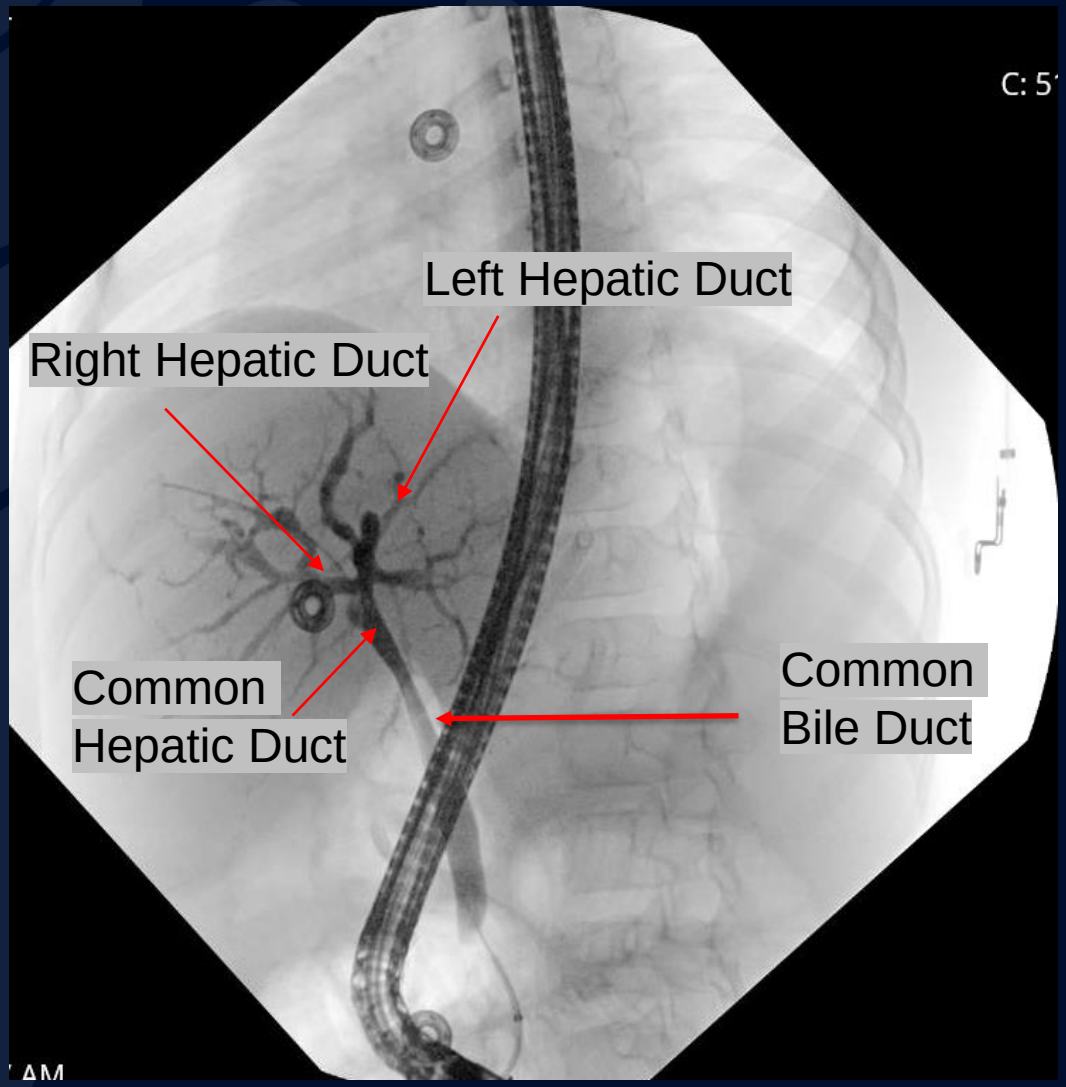
ERCP



No filling defects or stenosis within the CBD

Scattered areas of cystic dilatation of the intrahepatic biliary ducts

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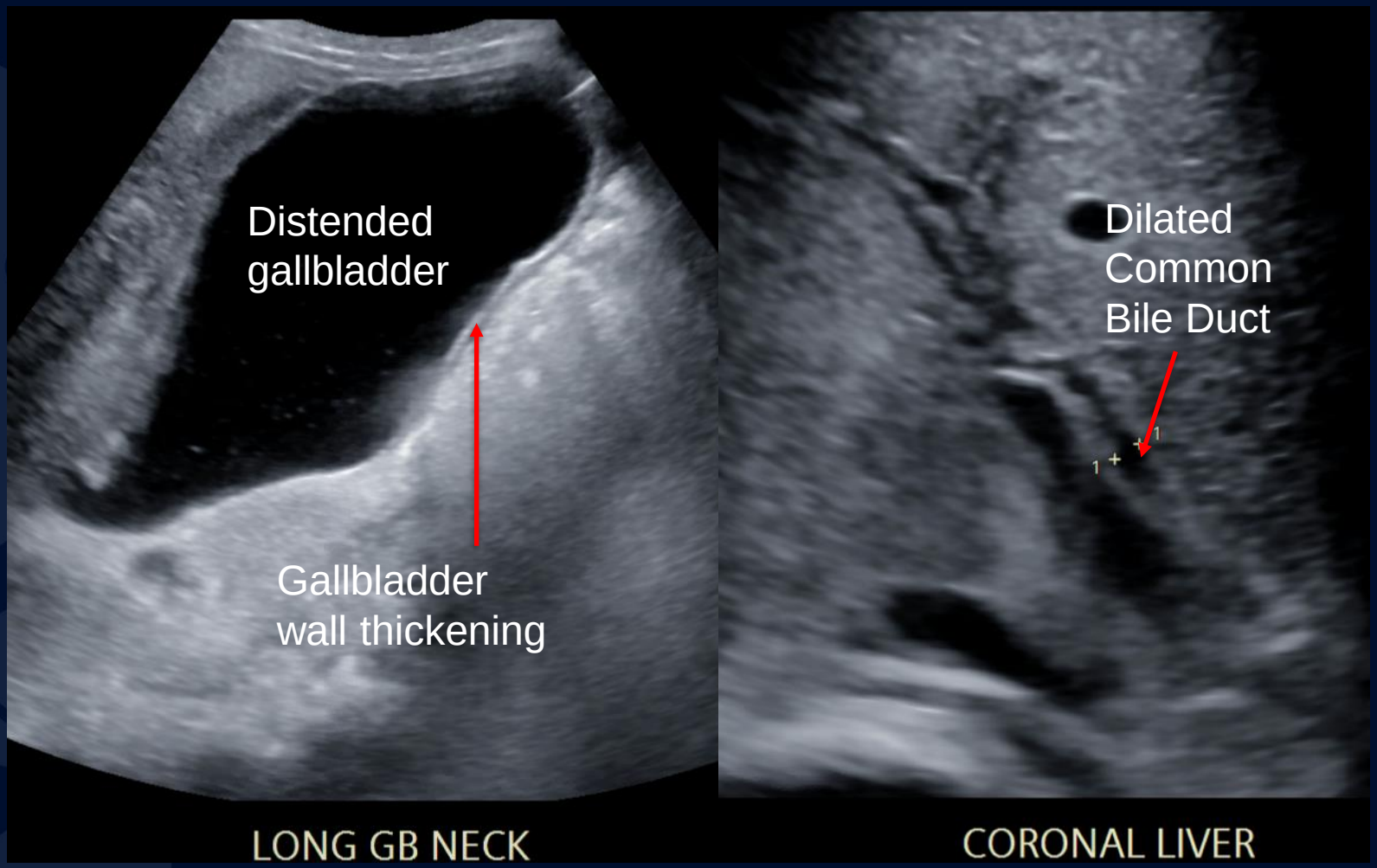
Ultrasound



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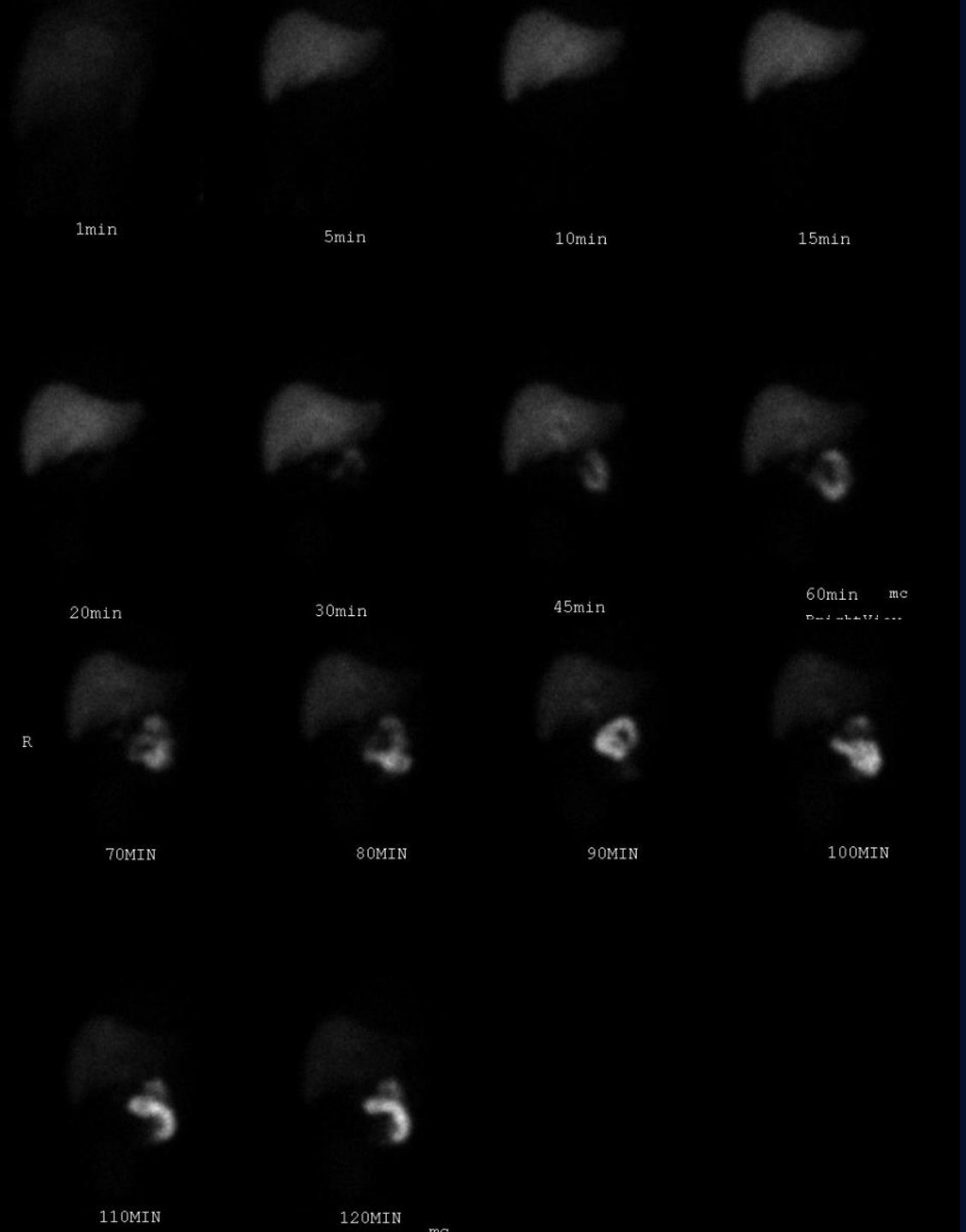

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Absent gallbladder visualization

Abnormal gallbladder emptying which supports suspicion of acalculous cholecystitis.

However, in setting of ERCP, can be a false positive due to preferential flow of contrast across the sphincterotomy

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Caroli Syndrome Pathophysiology

- Defective proteins in liver / kidney expressed on primary cilia and centrosome complex of renal tubule cells and cholangiocytes
- Pathogenesis of the intrahepatic ductal dilatation and hepatic fibrosis appears to be related to an arrest or derangement in remodeling of the ductal plate of the larger intrahepatic bile ducts during development
  - Caroli disease: abnormal remodeling occurs in early period of embryogenesis
  - Caroli syndrome: abnormal remodeling occurs later in embryogenesis→ resulting in abnormalities in the peripheral biliary ramifications (the intralobular bile ducts)
- Associated with autosomal recessive polycystic kidney disease (ARPKD) caused by pathogenic variants in the PKHD1(polycystic kidney and hepatic disease) gene
  - Expressed primarily in kidneys with lower levels in the liver, pancreas, lungs

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Imaging Findings

- Abdominal imaging (ultrasonography, CT scan, magnetic resonance cholangiography)
  - Bile duct ectasia and irregular, cystic dilation of the large proximal intrahepatic bile ducts with a normal common bile duct
  - “Central dot sign," defined as a small foci of contrast enhancement within dilated intrahepatic ducts, can be seen on CT or magnetic resonance imaging
- The disease may be limited to one lobe of the liver, commonly the left lobe
- Imaging can also show renal features of polycystic kidney disease

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References

- Suchy, Frederick J. “Caroli Disease.” *UpToDate*, https://www.uptodate.com/contents/caroli-disease.
- Sugiyama M, Izumisato Y, Abe N, et al. Cavernous pancreatic ductal ectasia with smooth muscle proliferation causing recurrent acute pancreatitis. *Int J Pancreatol*. 2001;29(2):99-105. doi:10.1385/IJGC:29:2:099
- Gallagher AR, Esquivel EL, Briere TS, Tian X, Mitobe M, Menezes LF, Markowitz GS, Jain D, Onuchic LF, Somlo S. Biliary and pancreatic dysgenesis in mice harboring a mutation in Pkhd1. Am J Pathol. 2008 Feb;172(2):417-29. doi: 10.2353/ajpath.2008.070381. Epub 2008 Jan 17. PMID: 18202188; PMCID: PMC2312372.

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