AMSER Case of the Month March 2025

2 year old male with vomiting

Jacob Rachwal OMS-III, PCOM

Dr. Ross Myers MD, Pediatric Radiology

Lehigh Valley Health Network

Patient Presentation

- HPI: 2 year old male presents to the ED for six episodes of vomiting in one day, decreased appetite, and decreased urination. He was at baseline before he started vomiting. Denies any apparent pain, diarrhea, cough, fever, or decreased responsiveness.
- PMH: Patient has a past medical history of malnutrition with 3 months of continuous NG tube feeds at 8 months old.
- Vitals: BP 106/73, HR 134, RR 26, temp 99.1 F
- Family History: No known history of GI diseases
- Medications: Acetaminophen 100mg

AMSER

Pertinent Labs

- Initial work up:

Liver Function Test	Level at Initial Work Up
AST	92 (elevated)
ALT	111 (elevated)
Alkaline Phosphatase	339 (elevated)
Total Bilirubin	0.4 (normal)

- Additional labs included elevated lipase (989) and amylase (179)
- Elevated hemoglobin/hematocrit (12.9/39.5), WBC (16.9), ANC (14.2), and platelets (446).
- Urinalysis was positive for proteins and ketones.

AMSER

Continued hospital course

- The patient was transferred to another ED for further management. Workup the following day included an abdominal ultrasound that showed findings of pancreatitis with a mildly dilated pancreatic duct up to 3 mm. It also showed a moderately dilated common bile duct up to 13 mm, sludge in distal common bile duct, and moderately distended gallbladder with minimal wall thickening and trace pericholecystic fluid.
- Further labs:

Liver Function Test	Level at Initial Work Up	Level One Day Later
AST	92 (elevated)	**223** (elevated)
ALT	111 (elevated)	**144** (elevated)
Alkaline Phosphatase	339 (elevated)	255 (normal)
Total Bilirubin	0.4 (normal)	**1.7** (elevated)
Direct Bilirubin	Not Measured	**1.3** (elevated)
GGT	Not Measured	206

AMSER

What Imaging Should We Order?

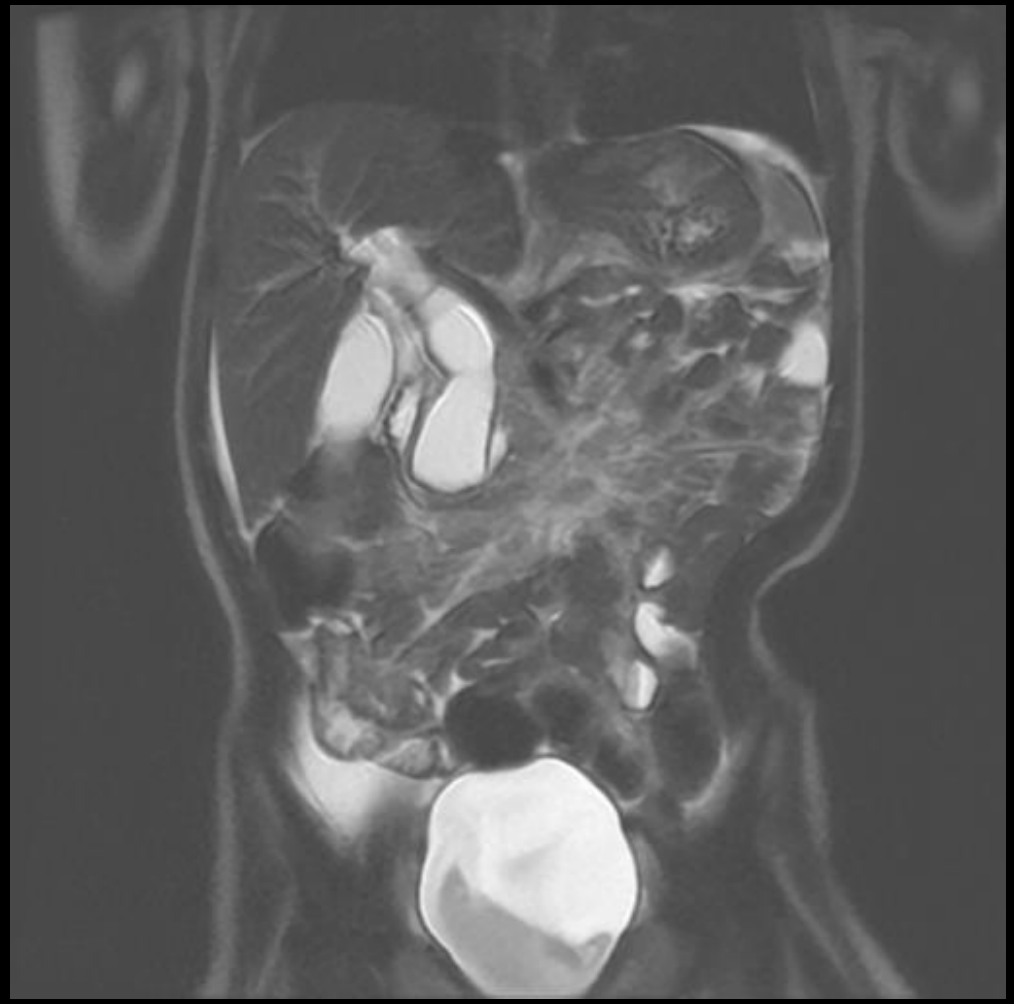
Select the applicable ACR Appropriateness Criteria

Abnormal liver function tests, hyperbilirubinemia, acute cholestasis, conjugated, initial imaging	3196533	US abdomen	0 mSv O	0 mSv [ped] O	Usually appropriate
		MRI abdomen without and with IV contrast with MRCP	0 mSv O	0 mSv [ped] O	Usually appropriate
		MRI abdomen without IV contrast with MRCP	0 mSv O	0 mSv [ped] O	Usually appropriate
		CT abdomen and pelvis with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually appropriate
		CT abdomen and pelvis without IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	May be appropriate
		US abdomen with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		US duplex Doppler abdomen	0 mSv O	0 mSv [ped] O	Usually not appropriate
		US shear wave elastography abdomen	0 mSv O	0 mSv [ped] O	Usually not appropriate
		MR elastography abdomen	0 mSv O	0 mSv [ped] O	Usually not appropriate
		CT abdomen and pelvis without and with IV contrast	10-30 mSv ☢☢☢☢	10-30 mSv [ped] ☢☢☢☢☢	Usually not appropriate

This imaging modality was ordered by the hospital physician

AMSER

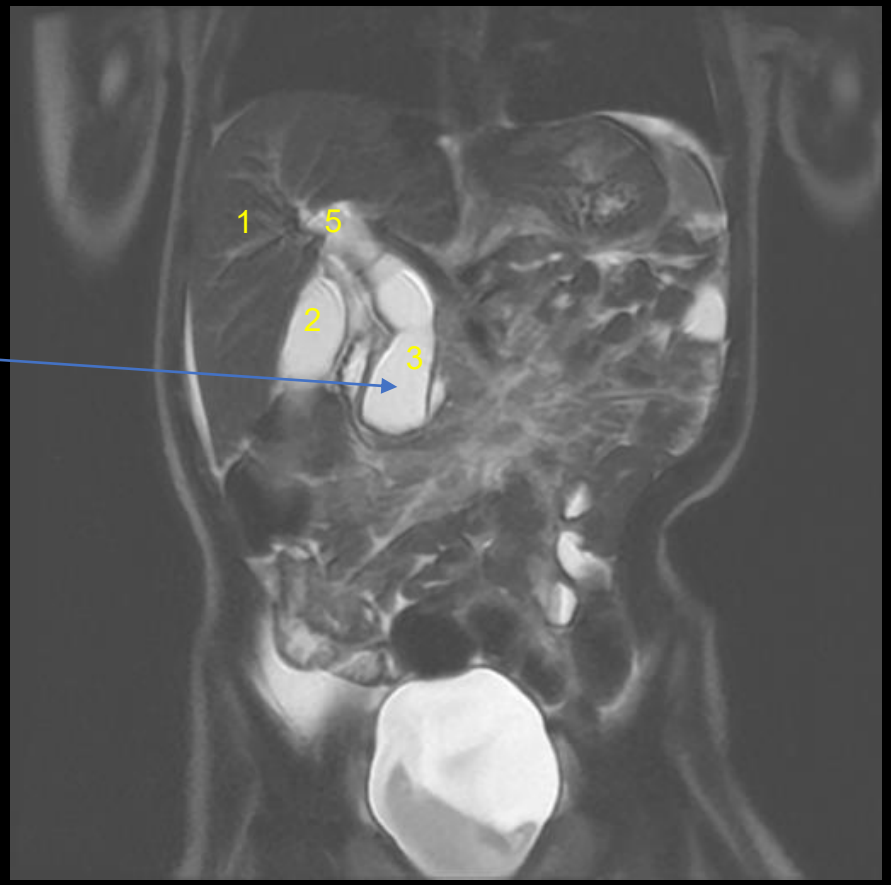
Findings (unlabeled)

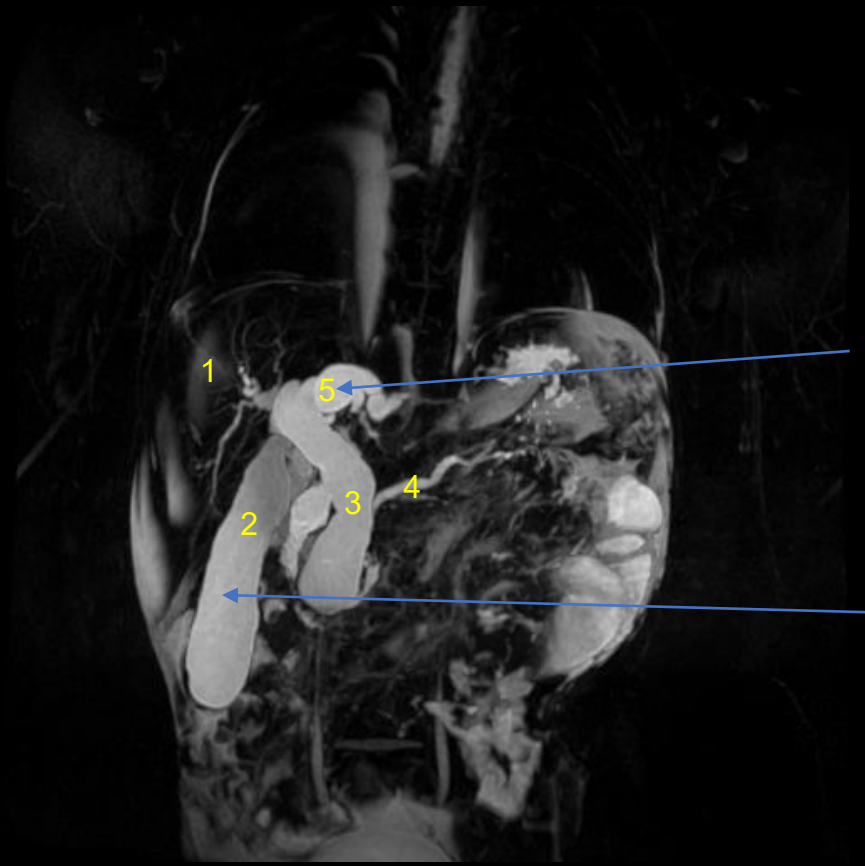


1 = Liver
2 = Gallbladder
3 = Common Bile duct
4 = Main pancreatic duct
5 = Intrahepatic ducts

Findings (labeled)

Extensive varicoid dilatation of the common bile duct



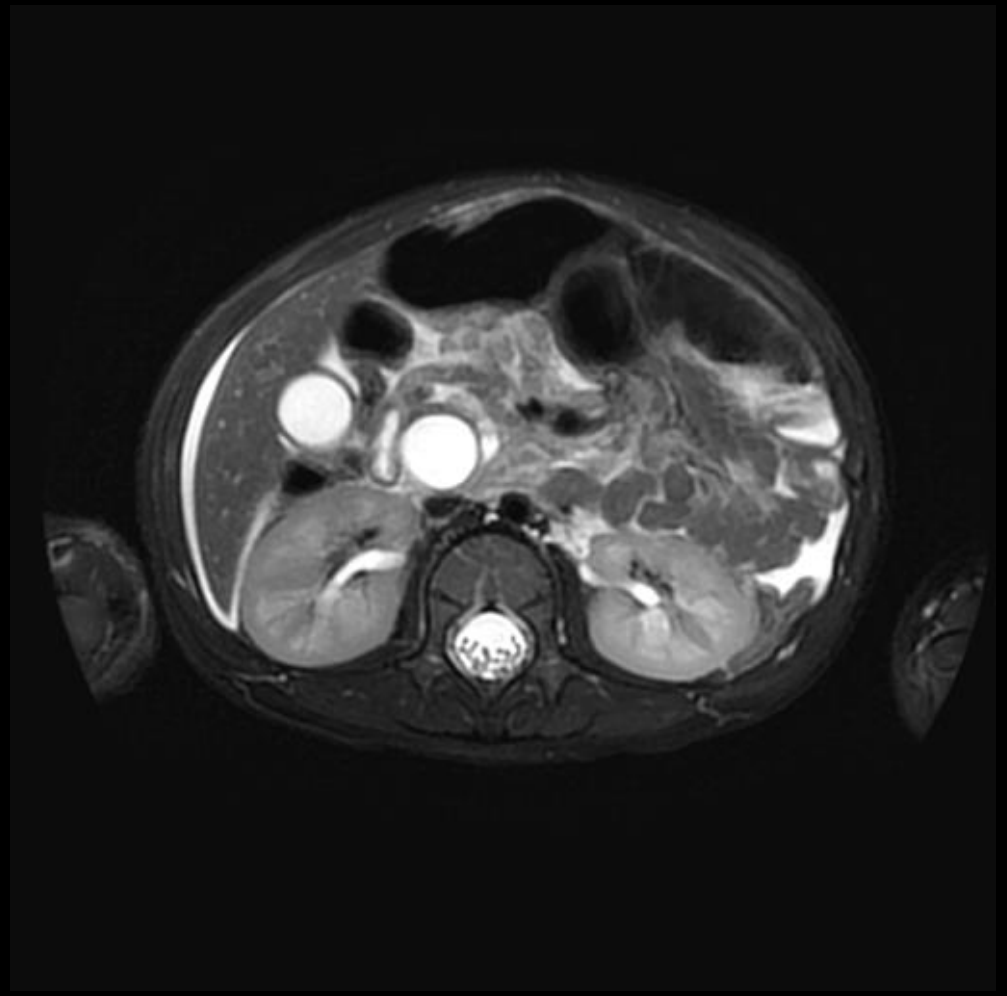


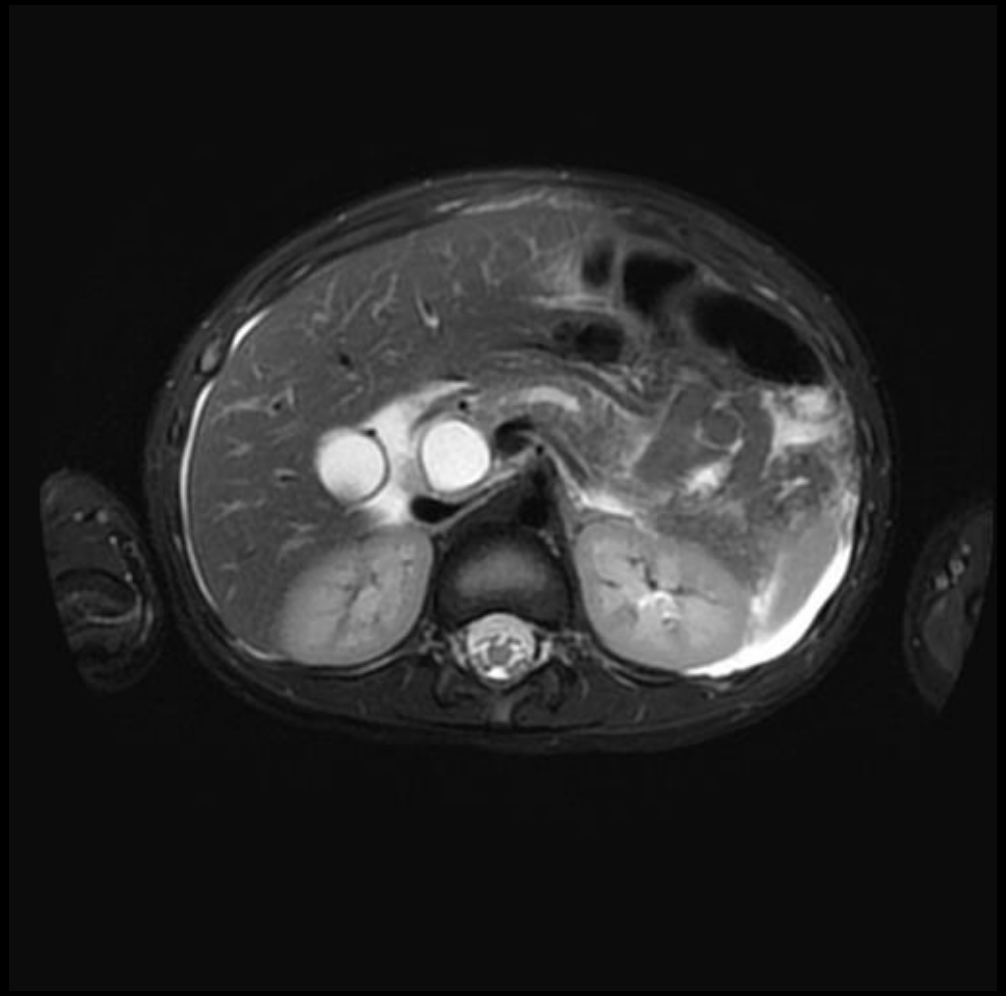
Extensive varicoid dilatation of the intrahepatic ducts

Physiologic distension of the gallbladder

AMSER

Findings: (unlabeled)



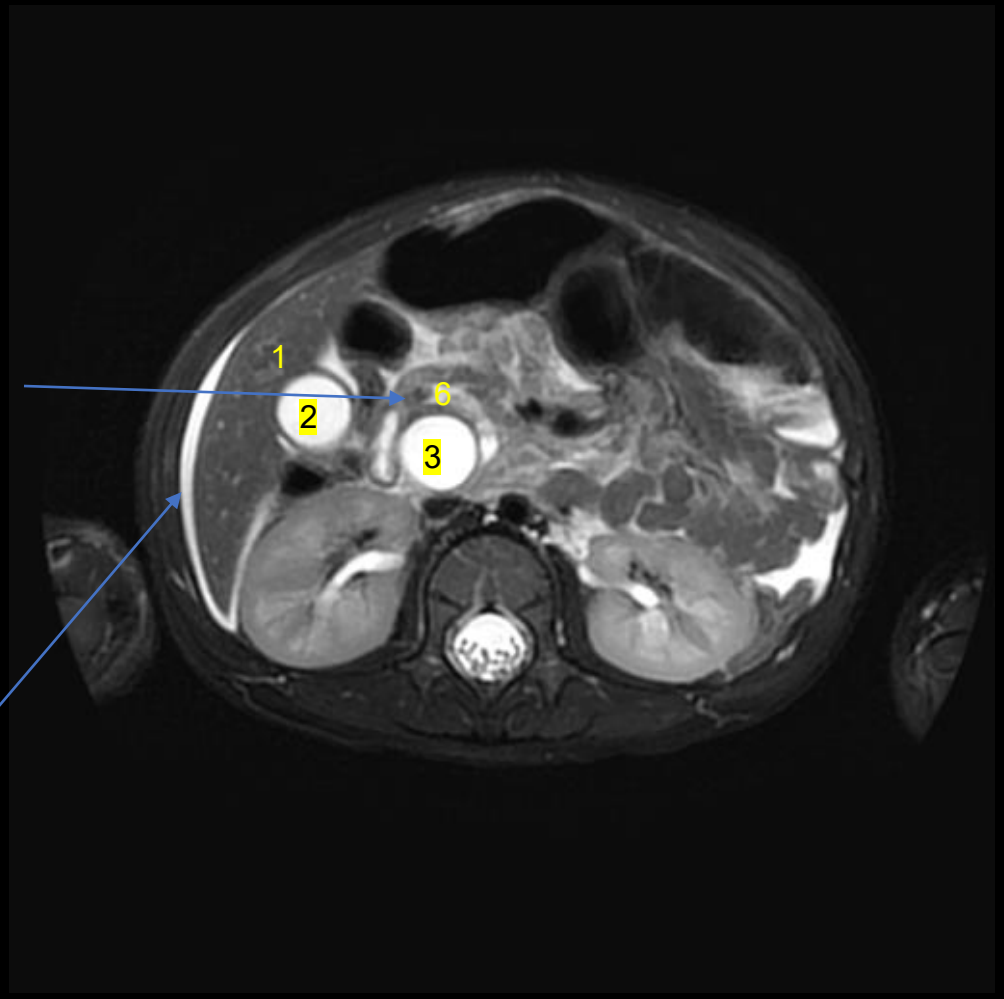


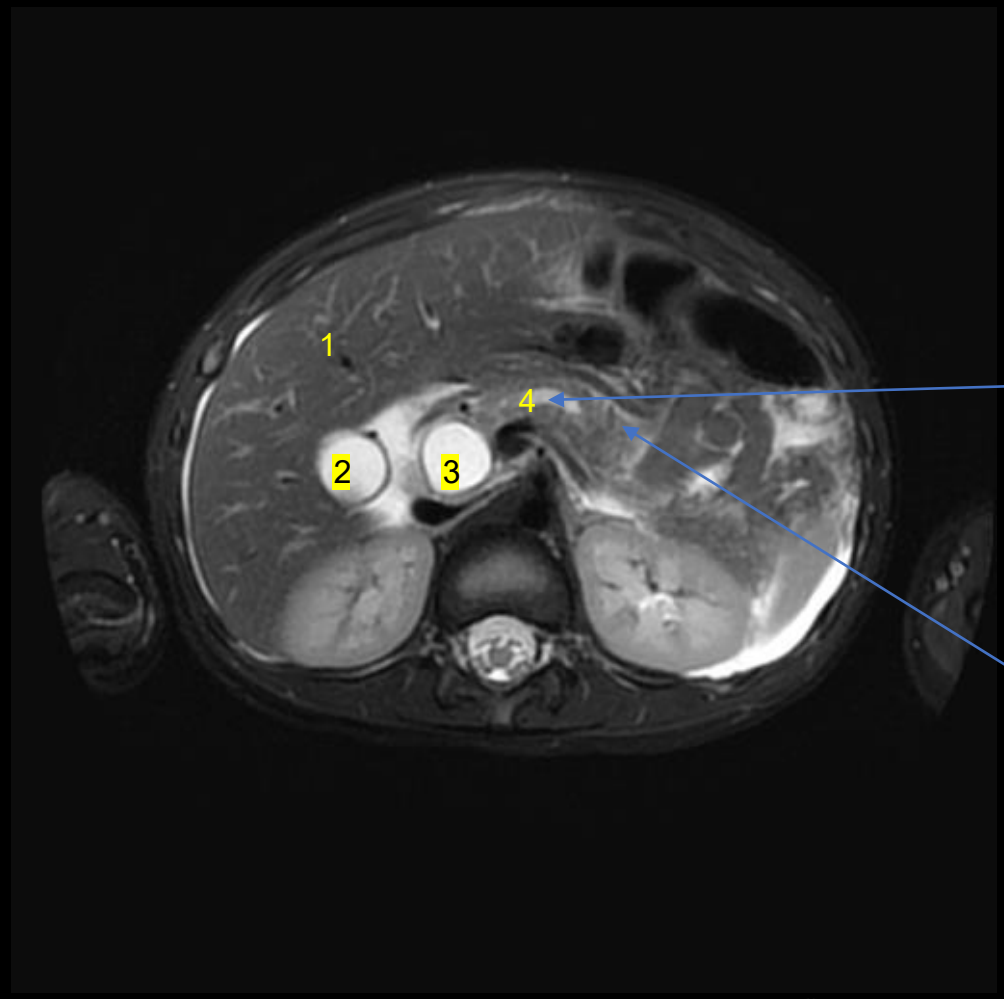
1 = Liver
2 = Gallbladder
3 = Common bile duct
4 = Main pancreatic duct
5 = Intrahepatic ducts
6 = Accessory duct of Santorini

Findings: (labeled)

Moderately dilated accessory duct of Santorini with concurrent 0.5cm signal void

Ascites





Diffuse moderate main pancreatic ductal dilatation

Pancreatic parenchymal edema

AMSER

Final Dx:

Choledochal cyst

- This patient has a choledochal cyst that is likely congenital. It is a type IVa choledochal cyst using the Todani classification system due to the presence of both intrahepatic and extrahepatic biliary ductal dilatation

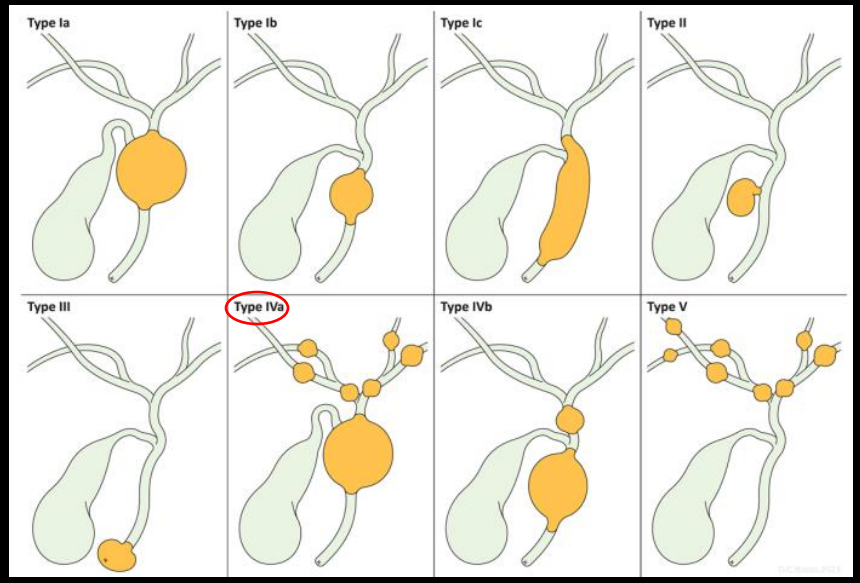


Figure 1. Todani classification system for choledochal cysts [1]

Case Discussion

- Choledochal cysts are extrahepatic and/or intrahepatic cystic dilatation of the biliary tree
- Congenital choledochal cysts are one cause of direct hyperbilirubinemia in pediatric patients due to obstructive jaundice. [2] Pancreatitis can occur due to stone and protein formation in the distal common bile duct and pancreatic duct. Ascending cholangitis is another presentation. [1]
- Classic triad of abdominal pain, jaundice and a palpable abdominal mass occurs in less than 20% of patients. [2]

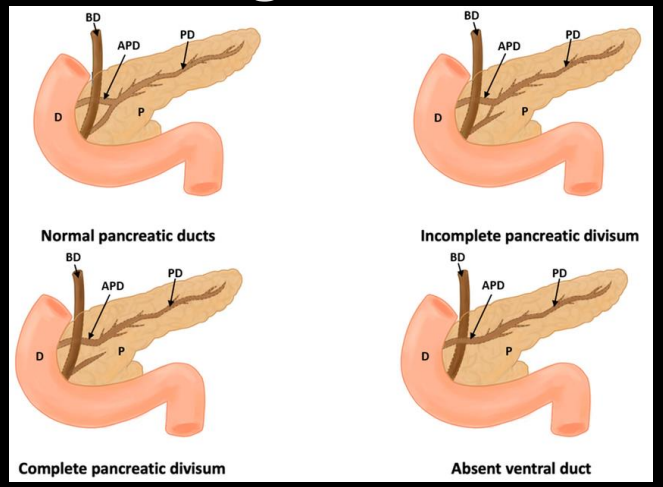
Case Discussion

- Pathophysiology: Unclear
- May be from pancreatobiliary maljunction which forms a common channel with subsequent reflux and pancreatic enzyme activation in the common bile duct which causes inflammation and intraductal pressure with cystic degeneration.[4]
- Diagnosis: Frequently detected with ultrasound, but MRCP is the optimal modality to evaluate the extent of involvement and the pancreaticobiliary junction.
- Treatment in most patients is total cyst excision with Roux-Y hepaticojejunostomy is the treatment of choice due to increased risk of cholangiocarcinoma.[5]

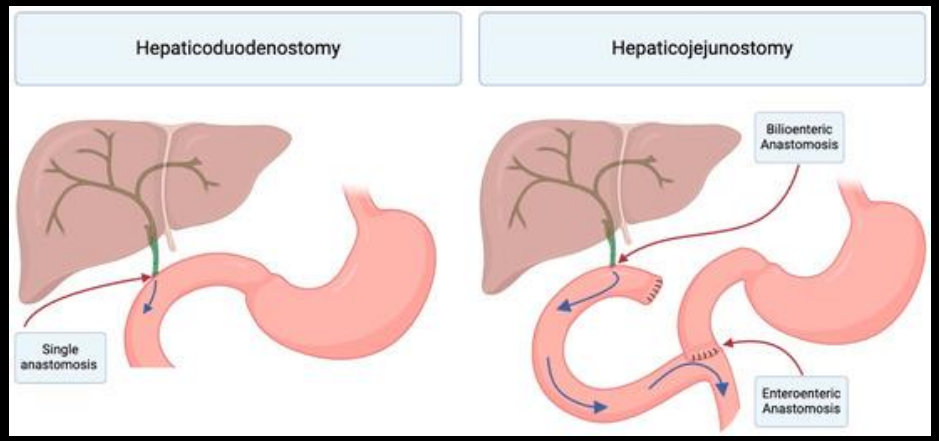
AMSER

Case Discussion

- Patient progress: Patient was discharged on day 7 with resolved symptoms of pancreatitis and transferred to a higher level of care where he had an ERCP which showed atrophic major papilla. However, there was drainage through the minor papilla which was stented. It also showed an incomplete pancreas divisum which may be related to the formation of his cysts.
- He will likely need choledochal cyst resection with Roux-en-Y hepaticojejunostomy. He will also need to be monitored yearly for cholangiocarcinoma.



Anatomical Variations in the pancreatic ductal system [6]



Biliary Reconstruction Methods after Choledochal Cyst Resection [7]

AMSER

References:

1. Lopes Vendrami C, Thorson DL, Borhani AA, et al. Imaging of Biliary Tree Abnormalities. *Radiographics*. 2024;44(8):e230174. doi:10.1148/rg.230174 .
2. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 2 of 3: Diagnosis. *Can J Surg*. 2009;52(6):506-511.
3. Ochiai K, Kaneko K, Kitagawa M, Ando H, Hayakawa T. Activated pancreatic enzyme and pancreatic stone protein (PSP/reg) in bile of patients with pancreaticobiliary maljunction/ choledochal cysts. *Dig Dis Sci*. 2004;49(11-12):1953-1956. doi:10.1007/s10620-004-9599-7.
4. Le L, Pham AV, Dessanti A. Congenital dilatation of extrahepatic bile ducts in children. Experience in the central hospital of Hue, Vietnam. *Eur J Pediatr Surg*. 2006;16(1):24-27. doi:10.1055/s-2005-873071.
5. Jabłońska B. Biliary cysts: etiology, diagnosis and management. *World J Gastroenterol*. 2012;18(35):4801-4810. doi:10.3748/wjg.v18.i35.4801.
6. Orellana-Donoso M, Milos-Brandenberg D, Benavente-Urtubia A, et al. Incidence and Clinical Implications of Anatomical Variations in the Pancreas and Its Ductal System: A Systematic Review and Meta-Analysis. *Life (Basel)*. 2023;13(8):1710. Published 2023 Aug 9. doi:10.3390/life13081710.
7. Iglesias N, Huerta CT, Lynn R, Perez EA. Biliary Reconstruction with Hepaticoduodenostomy Versus Hepaticojejunostomy After Choledochal Cyst Resection: A Narrative Review. *Journal of Clinical Medicine*. 2024; 13(21):6556. https://doi.org/10.3390/jcm13216556.

AMSER