Case 7729
Child with choledochal cyst presenting with episodes of vomitting and jaundice

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Section: Paediatric Radiology
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Patient: 2 year(s), female

Clinical History

A 2 year old girl presented to the emergency department with frequent episodes of vomiting and jaundice. Analytically, there was leucocytosis with normal neutrophil count, RCP of 5, 66 mg/dL and GGT 87 U/L. Colluria was also found.

Imaging Findings

A 2 year old girl presented to the emergency department with frequent episodes of vomiting and jaundice. Analytically, there was leucocytosis with normal neutrophil count, RCP of 5.66 mg/dL and GGT 87 U/L. Colluria was also found.
An abdominal ultrasound and Doppler-ultrasound were performed, which showed regular dilatation of the common bile duct, with a thin and regular wall, and without signs of an obstructive element either intra or extra-luminal. There was no cholelithiasis.
MR cholangiography showed a cystic fusiform dilatation of the main bile duct. The patient was submitted to surgical excision of the cyst with cholecistectomy and hepatico-jejunostomy, and recovered uneventfully.

Discussion

Choledochal cysts are rare congenital biliary tract anomalies which present as dilatation of the biliary tree, either all or part of the extra-hepatic or intra-hepatic ducts [1, 2]. Most often, choledochal cysts are diagnosed in childhood, however, in up to 20% of cysts are diagnosed in adults [2].
The clinical presentation is nonspecific, which often leads to a delay in diagnosis, in adults [3]. The triad of abdominal pain, a right upper quadrant mass, and jaundice is more prevalent in the paediatric population and is reported to occur in 2-38% of patients [2]. There is a 4:1 female predilection [4].

Todani classification of choledocal cysts (Fig 4):
Type I: these are cysts which involve only the extra-hepatic duct and result from an anomalous pancreaticobiliary union with formation of a long, frequently dilated common channel. They can be subdivided into type Ia, diffusely involving all of the extra-hepatic duct; Type Ib, which are characterised by focal dilatation of the extra-hepatic duct; and Type Ic, characterised by fusiform dilatation of the common bile duct

Todani Type II choledochal cysts are true diverticula of the extra-hepatic ducts [4].

Todani Type III choledochal cysts or choledochoceles, are characterised by segmental, intramural dilatation of the common bilde duct and manifest as focal dilatation of the intraduodenal segment of the distal common bile duct and may present with intermittent biliary colic, jaundice, and pancreatitis [4].

Todani Type IV choledochal cysts are by definition multiple and can involve both the intrahepatic and extrahepatic ducts: type IVa cysts involve both the extra and intra-hepatic ducts; and type IVb cysts are characterised by multiple saccular dilatations of the extra-hepatic ducts [4].

Todani Type V choledochal cysts or Caroli disease is a rare congenital, autosomal recessive disorder, characterised by cystic dilatation of the intra-hepatic bile ducts.
Clinically, it may manifest as recurrent bouts of cholangitis due to bile stasis, right upper auadrant abdominal pain, fever, and, more rarely, jaundice [5].
On imaging studies, there are intrahepatic saccular or fusiform dilated cystic structures of varying sizes which are seen to communicate with the biliary tree. The central dot sign, which is considered highly suggestive of Caroli disease results from enhancing portal branches surrounded by cystic alterations of the intra-hepatic ducts represented by an enhancing tiny dot in the middle of the dilated intra-hepatic ducts [6, 7].

The differential diagnosis includes primary sclerosing cholangitis, recurrent pyogenic cholangitis, autosomal dominant polycystic liver disease, biliary hamartomas, microabscess, biliary papillomatosis, and, occasionally, obstructive biliary dilatation
[8].
It is associated to other choledocal cysts and renal cystic disease such as medullary sponge kidney and tubular ectasia, and higher risk for cholangiocarcinoma [9].

Final Diagnosis

Type I choledocal cyst (Todani classification)

Figures

Figure 1 Abdominal ultrasound and doppler-ultrasound- Main bile duct dilatation (16.7 mm)

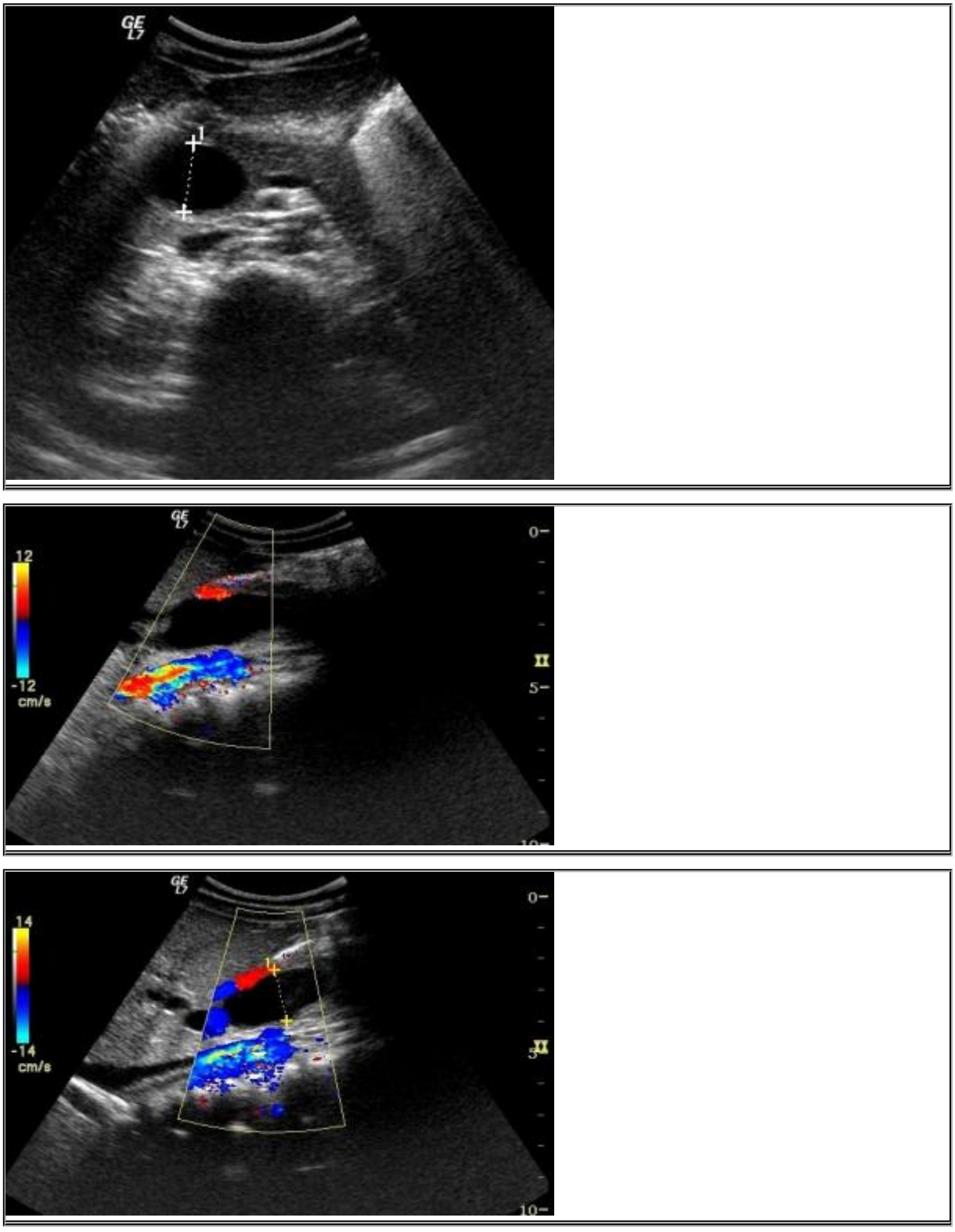
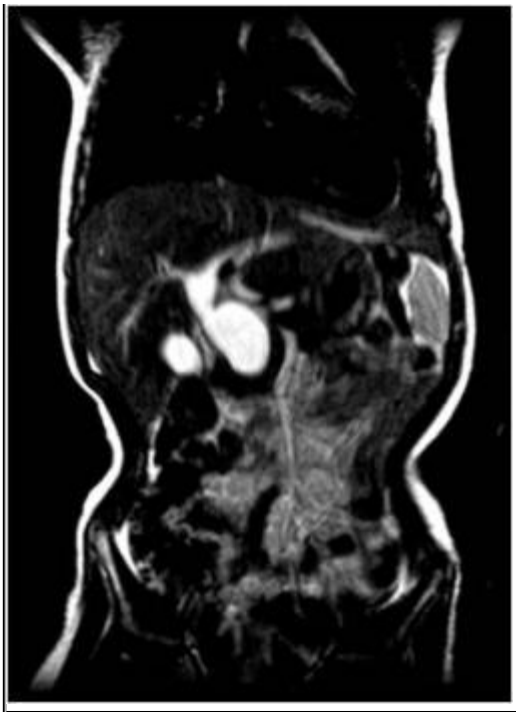
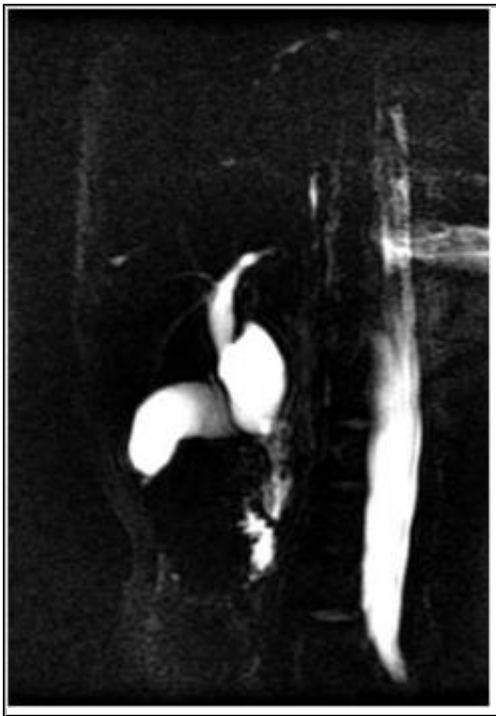


Figure 2 MRI- T2 Coronal



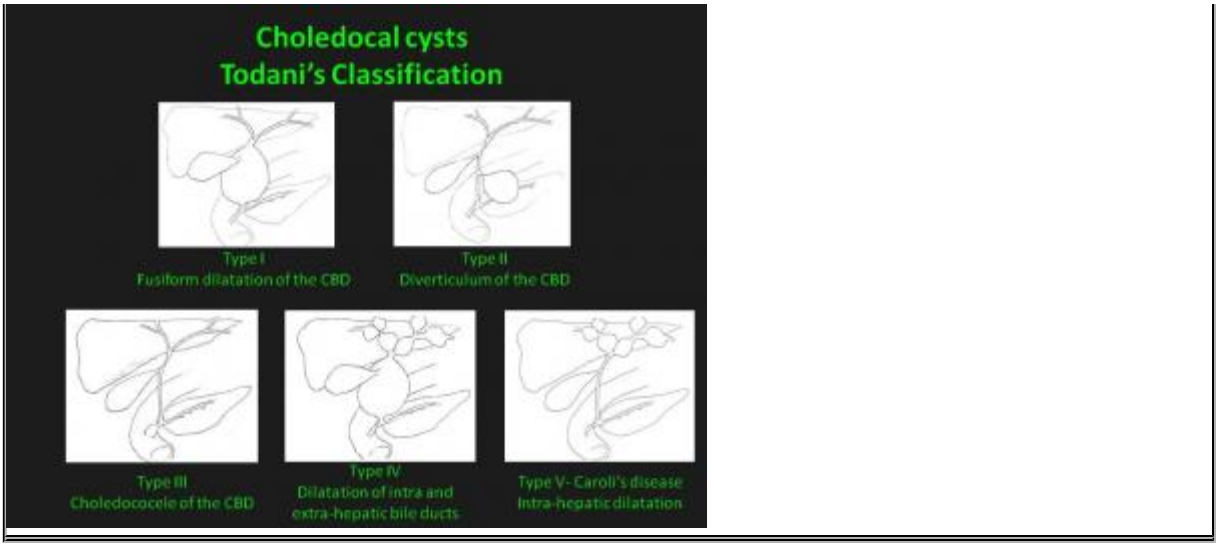
MR Cholangiography showed a cystic fusiform dilatation of the main bile duct (type I choledocal cyst).

Figure 3 MR Cholangiography



Fusiform dilatation of the main bile duct

Figure 4 Choledocal cysts. Todani's Classification



MeSH

Choledochal Cyst [C06.130.120.127]
A congenital anatomic malformation of a bile duct, including cystic dilatation of the extrahepatic bile duct or the large intrahepatic bile duct. Classification is based on the site and type of dilatation. Type I is most common.

References

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Citation

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