An exceedingly rare case of separated drainage of the cystic duct, the common bile duct and the main pancreatic duct documented on magnetic resonance cholangiopancreatography

Nicolò Brandi[1], Marta Fiscaletti[2], Matteo Renzulli[1]

[1] Department of Radiology, IRCCS Azienda Ospedaliero-Universitaria di Bologna, Via Albertoni 15, Bologna, Italy

[2] Radiology Units, Bellaria and Maggiore Hospitals, AUSL Bologna, Bologna, Italy

SUMMARY

Anatomical variations of the intra- and extra-hepatic biliary system are common, including those affecting the course and insertion point of the cystic duct. Adequate knowledge of such variations and an appropriate roadmap before any surgical, endoscopic or percutaneous procedure help in preventing associated iatrogenic complications. Magnetic resonance cholangiopancreatography (MRCP) can precisely delineate the anatomy of the biliary system preoperatively. We report the case of a 72-year-old female patient who presented with chronic right upper quadrant abdominal pain of 6 months duration, which had acutely worsened over the previous 2 weeks, and no other comorbidities. Blood tests were normal. Abdominal ultrasound revealed a slight dilatation of the common hepatic duct (10 mm), but no gallstones or sludge were demonstrated. MRCP excluded choledocholithiasis but revealed a cystic duct coursing medial to the common bile duct before anastomosing at the level of the ampulla of Vater. Trifurcation (Type 2 variant) of the intra-hepatic bile system was also observed. To the best of the authors' knowledge, this is the very first report to clearly document this exceptionally rare anatomic cystic duct anomaly on MRCP.

Key words: Biliary tract – Cystic duct – Cholangiopancreatography – Magnetic resonance

ABBREVIATIONS:

CBD: common bile duct
CD: cystic duct
CHD: common hepatic duct
CT: computed tomography
EHBD: extra-hepatic biliary ducts
ERCP: endoscopic retrograde cholangiography
FIESTA: fast imaging employing steady-state acquisition
FSEGR: fast spoiled gradient echo
HR: high resolution
IHBD: intra-hepatic biliary ducts
MRCP: magnetic resonance cholangiopancreatography
SSFSE: single shot fast spin echo
US: ultrasound

Corresponding author:
Nicolò Brandi. Department of Radiology, IRCCS Azienda Ospedaliero-Universitaria di Bologna, Via Albertoni 15, 40138 Bologna, Italia. Phone: +390512142958, Fax: +390512142805. E-mail: nicolo.brandi2@unibo.it

Submitted: June 2, 2023. **Accepted:** July 18, 2023

https://doi.org/10.52083/JHOF9715

INTRODUCTION

Anomalies of the biliary ductal system have been appreciated by physicians and anatomists for centuries and are common findings, possibly involving both the intra-hepatic biliary ducts (IHBD) and the extra-hepatic biliary ducts (EHBD).

Normally, the cystic duct (CD) joins the common hepatic duct (CHD) from a right lateral position approximately halfway between the porta hepatis and the ampulla of Vater. The significance of a low CD union was first described by Eisendrath in 1918, who noted that such anatomy might predispose to inadvertent bile duct injury during cholecystectomy (Eisendrath, 1918). Reports concerning the further clinical significance of this anatomic anomaly are relatively rare, but they observed an association with gallstone pancreatitis, Mirrizi syndrome, gallbladder cancer, and cystic dilatation of the biliary duct (Kubota et al., 1993; Uetsuji et al., 1993).

Despite a medial insertion of CD is reported in about 16-18% of patients, its low insertion has been observed as more infrequent and has been described in only 6-16%; in addition, the combination of these two anatomical variations is even more uncommon, presenting merely in 4-5.5% of cases (Gündüz et al., 2021; Sarawagi et al., 2016; Shaw et al., 1993; Taourel et al., 1996; Tsitouridis et al., 2007; Turner & Fulcher, 2001).

A medial and extremely low (intraduodenal) insertion of CD is an exceedingly rare finding, with percentages ranging from 1.66% in cadaveric studies (Sangameswaran, 2021) to 2% in cholangiopancreatographic studies (Garg et al., 2022), and only very few cases reported in the literature. Moreover, evidence of this singular configuration is supported only by endoscopic retrograde cholangiopancreatography (ERCP) images (Dodda et al., 1998; Turner & Fulcher, 2001). The present case is a unique case of medial and extremely low (intraduodenal) insertion of CD and, to the best of the authors' knowledge, the very first report to clearly document this exceptionally rare anatomic anomaly on magnetic resonance cholangiopancreatography (MRCP).

CASE REPORT

A 72-year-old female reported to our Institution with chronic right upper quadrant abdominal pain of 6 months duration, which had acutely worsened over the previous 2 weeks. She had no other comorbidities. The patient had been empirically treated with ursodeoxycholic acid prescribed by her primary care physician without improvement. Blood tests were normal, with lipase levels within normal limits and no leukocytosis. In the suspicion of biliary stones, an abdominal ultrasound (US) was requested. US revealed a slight dilatation of the CHD (10 mm), but no gallstones or sludge were demonstrated; furthermore, the gallbladder and the intrahepatic biliary ducts did not reveal any obvious abnormality. To definitively exclude choledocholithiasis and gallstones, an MRCP was then performed.

The MRCP was performed using a 1.5T MRI superconductive scanner (HDX-t Signa; General Electric®, Milwaukee, WI, USA) after the administration of one glass of pineapple juice to reduce the signal hyperintensity of gastro-enteric fluids; in particular, the MRCP protocol included the following sequences: axial fast spoiled gradient echo (FSEGR) T1-weighted images (including both in- and out-of-phase), axial single-shot fast spin echo (SSFSE) T2-weighted images, high resolution (HR) thick slab images and 3D fast imaging employing steady-state acquisition (FIESTA) images. The MRCP excluded the presence of any stones in the biliary system and in the gallbladder, but revealed a very rare cystic duct anomaly, demonstrating a CD coursing parallel and medial to the common bile duct (CBD) before anastomosing at the level of the ampulla of Vater; therefore, three distinct bile draining ducts were noted separately arising from the ampulla: the CD, the CBD and the main pancreatic duct (Fig. 1A-C). In addition, an anatomical variation of the IHBD was also observed, with a common confluence of the right posterior duct, the right anterior duct and the left hepatic duct (i.e., trifurcation or Type 2) (Fig. 1D). Furthermore, the gallbladder showed the folding of its fundus back upon the body, configuring a "Phrygian cap" anomaly, and demonstrated the presence of fundal adenomyomatosis (Fig. 1D-E).

COMMENTS

The CD normally joins the CHD approximately halfway between the porta hepatis and the ampulla of Vater from a right lateral position.

However, anatomical anomalies of the CD might occur in 18-23% of patients (Shaw et al., 1993) due to variations in the length, course and/or pattern of entry of the CD into the EHBT. According to the available literature, despite the level of insertion of the CD generally occurring in the middle one-third of the EHBT (51-88% of cases), the union may also occur in both the proximal and distal one-third (6-35% and 6-16%, respectively). Moreover, when it inserts distally, the CD may run parallel with the CHD for a long distance before fusion (12-33%) or, occasionally, even spiral around it (10%). In some rare cases of abnormal proximal insertion, the CD may drain directly into the right or left hepatic duct. As concerns the pattern of entry, the CD enters the CBD from the lateral aspect in most cases (37-68%), whereas a medial insertion after crossing the front or from behind is less common (16-18%); in some cases, the CD may present an anterior or posterior insertion (13-17% and 19-26%, respectively). Other unusual and rarer anomalies include double CDs and an absent CD with the gallbladder often emptying via a different pathway (Garg et al., 2022; Gündüz et al., 2021; Mortelé and Ros, 2001; Pina et al., 2015; Turner and Fulcher, 2001).

The possible anatomical variants of the CD are therefore extremely various and most probably depend on both the timing of the process of separation of pars hepatica (which will become the liver, the IHBD system and the CHD) from pars cystica (which will form the CD, the CBD and the

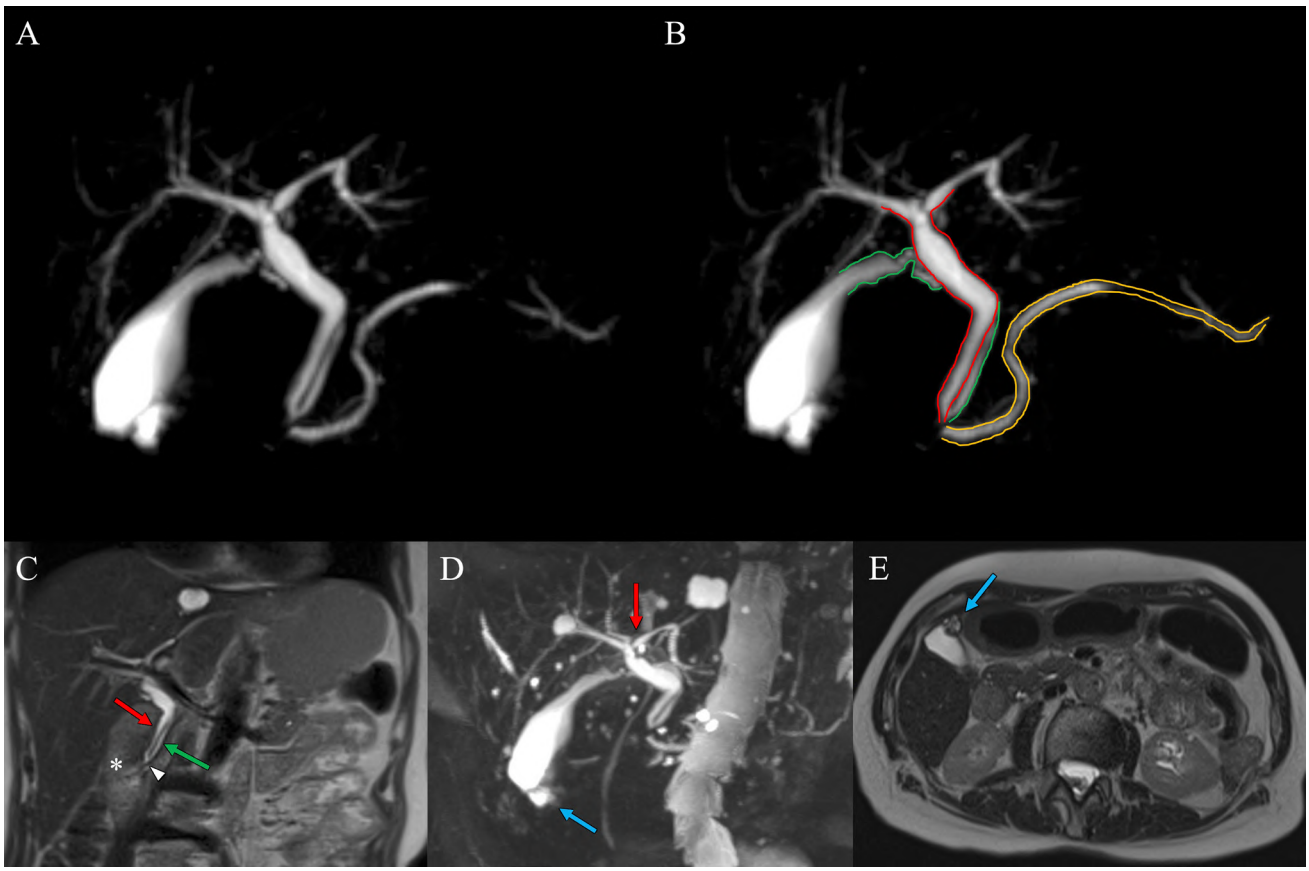

Fig. 1.- Coronal MRCP HR images (**A** and **B**) demonstrating a cystic duct (CD, green outline in B) coursing parallel and medial to the common bile duct (CBD, red outline in B) before anastomosing at the level of the ampulla of Vater with the main pancreatic duct (yellow outline in B); no stones were detected in both the gallbladder and the extra-hepatic biliary system. Coronal T2-w image (**C**) demonstrating the confluence of the CBD (red arrow) and the CD (red arrow) in the ampulla of Vater (white arrowhead) and showing the second portion of the duodenum (white asterisk). Oriented MRCP HR image obtained after maximum intensity projection (MIP) reconstruction (**D**) of the same patient demonstrating the concomitant presence of Type 2 variant (trifurcation) of the intra-hepatic bile duct (red arrow) and "Phrygian cap" anomaly of the gallbladder fundus (blue arrow). Axial T2-w image (**E**) demonstrating the presence of fundal adenomyomatosis in the gallbladder (blue arrow).

gallbladder), and the grade of rotation of the CD itself induced by the rotation of the duodenum during the fetal stages (Lamah et al., 2001; Tan and Moscoso, 1994). Whether these anatomical variations are due to simple stochastic events or rather represent the outcome of delays and/or alterations in the "normal" rates of differential growth in the different embryonic duct structures is still unknown. Moreover, since the EHBD and the IHBD maintain luminal continuity from the very start of organogenesis (Roskams and Desmet, 2008), it cannot be excluded that variations in the configuration of one system may induce or, at least, contribute to a concomitant alteration in the other system, causing some sort of "ripple effect" on the following morphogenetic phases. Nonetheless, recent evidence seems to support well-distinct mechanisms regulating the development of both the EHBD and the IHBD systems, suggesting that the molecular pathways involved in the development of the EHBD are more closely related to the formation of the duodenum and the pancreas (Zong and Stanger, 2011). Therefore, further studies are warranted to elucidate the mechanisms responsible for variations in the remodeling process of the biliary system during embryogenesis.

Variations in the course and insertion point of the CD are common in clinical practice and they typically remain asymptomatic throughout life. However, these anomalies still demand considerable attention during diagnostic investigations and interventional and surgical procedures since they might be the source of complications. For example, a CD with an extremely low and medial insertion has the tendency to overlie the CBD and thus be misidentified prior to interventional procedures, thus increasing the risk of inadvertent ERCP stent malposition during stone retrieval (George et al., 2009). In addition, in the case of an extremely low union, the CD and the CBD may be joined by fibrous tissue, thus making clamping the CD difficult or resulting in the inadvertent iatrogenic injury of the CD. Finally, the presence of an unusually long CD remnant after cholecystectomy or living donor liver transplantations has been associated with inflammatory changes and the formation of calculi (Turner and Fulcher, 2001).

Therefore, noninvasive techniques that can precisely delineate the anatomy of the biliary tract preoperatively could be of clinical value. In most cases, the normal-caliber CD is not seen on US or computed tomography (CT). Conversely, MRCP proved to be an accurate non-invasive imaging modality for mapping the CD anatomy and detecting aberrant ductal anatomy, relying on the high signal intensity of fluid-containing structures in T2-weighted images (Taourel et al., 1996). This technique allows to obtain high-resolution cross-sectional imaging, with both two- and three-dimensional projection, and its results are comparable to the more invasive ERCP and intraoperative cholangiograms (Hekimoglu et al., 2008; Xu et al., 2013). Moreover, MRCP is quick, is not associated with radiation exposure and does not require the administration of contrast media, needing only a simple glass of juice to evaluate the anatomy of both the intra- and extrahepatic biliary ducts (Renzulli et al., 2022). Finally, it allows the synchronous evaluation of the IHBD, whose anatomical variations are often associated with variations of the downstream biliary system, as was demonstrated also in the present case (Renzulli et al., 2023).

Despite the potential surgical complications associated with these rare congenital malformations, CD aberrations have been demonstrated to predispose to several pathologic pancreaticobiliary conditions. For example, a low union of the CD has previously been associated with a higher risk of pancreatitis and even periampullary cancer, and this association was ascribed to an anatomy-induced pressure process leading to chemical/bilious irritation (Muraki et al., 2020); this association is probably supported also by the shorter and lesser exposure of the biliary epithelium to the protective or dilutional effect of peri-biliary mucus glands in the upper biliary tree which happens in the presence of a low union (Dodda et al., 1998). In addition, previous studies have found an association between low insertion of the CD and choledocholithiasis (Renzulli et al., 2021) which can be due to increased retrograde pressure in the CD leading to both bile stagnation and impaired gallbladder emptying. In the present case, however, no calculi were detected on both US and MRCP. Despite

being contradictory to previous reports, the absence of biliary stones in the present case may be explained by differences in terms of biochemical and clinical contributors to stone formation. At the same time, the chronic abdominal pain reported by the patient could be attributed to unusual bile reflux into the CD or the pancreatic duct, likely favored by the relative proximity of these canals and the increased intra-luminal pressure at the level of the anastomosis. This hypothesis could also justify the evidence of the slightly wider CHD diameter reported on US and MRCP.

CONCLUSION

A CD coursing parallel and medial to the CBD before anastomosing at the level of the ampulla of Vater is an extremely rare anatomical variant of the biliary system that, if unrecognized, may complicate both surgical and interventional procedures. In an era of ever-increasing laparoscopic, endoscopic and percutaneous procedures of the hepatobiliary system, it is extremely important to have adequate knowledge of such variations before the actual procedure is performed. MRCP is pivotal to correctly identifying anatomic variants of both the IHBD and the EHBD, whose association is not unusual.

REFERENCES

DODDA G, BROWN, RD, O'NEIL HK, VENU RP (1998) Cystic duct insertion at ampulla as a cause for acute recurrent pancreatitis. *Gastrointest Endosc*, 47(2): 181-183.

EISENDRATH DN (1918) Anomalies of the bile ducts and blood vessels as the cause of accidents in biliary surgery. *JAMA*, 71(11): 864-867.

GARG S, DUTTA U, CHALUVASHETTY SB, KUMAR KH, KARLA N, SAHINI D, AGGARWAL A (2022) The anatomy of the cystic duct and its association with cholelithiasis: MR cholangiopancreatographic study. *Clin Anat*, 35(7): 847-854.

GEORGE RA, DEBNATH J, SINGH K, SATIJA L, BHARGAVA S, VAIDYA A (2009) Low insertion of a cystic duct into the common bile duct as a cause for a malpositioned biliary stent: demonstration with multidetector computed tomography. *Singapore Med J*, 50(7): e243-e246.

GÜNDÜZ N, DOĞAN MB, ALACAGÖZ M, YAĞBASAN M, PERÇEM ORHAN SÖYLEMEZ U, ATALAY B (2021) Anatomical variations of cystic duct insertion and their relationship with choledocholithiasis: an MRCP study. *Egypt J Radiol Nucl Med*, 52: 202.

HEKIMOGLU K, USTUNDAG Y, DUSAK A, ERDEM Z, KARADEMIR B, AYDEMIR S, GUNDOGDU S (2008) MRCP vs. ERCP in the evaluation of biliary pathologies review of current literature. *J Diag Dis*, 9(3): 162-169.

KUBOTA Y, YAMAGUCHI T, TANI K, TAKAOKA M, FUJIMURA K, OGURA M, YAMAMOTO S, MIZUNO T, INOUE K (1993) Anatomical variation of pancreatobiliary ducts in biliary stone diseases. *Abdom Imaging*, 18(2): 145-149.

LAMAH M, KARANJIA ND, DICKSON GH (2001) Anatomical variations of the extrahepatic biliary tree review of the world literature. *Clin Anat*, 14(3): 167-172.

MORTELÉ KJ, ROS PR (2001) Anatomic variants of the biliary tree: MR cholangiographic findings and clinical applications. *Am J Roentgenol*, 177(2): 389-394.

MURAKI T, REID MD, PEHLIVANOGLU B, GONZALEZ RS, SEKHAR A, MEMIS B, XUE Y, CHENG J, JANG KT, MITTAL P, CARDONA K, KOOBY DA, MAITHEL S, SARMIENTO JM, EL-RAYES B, LOMBERK G, URRUTIA RA, CHRISTIANS K, TSAI S, EVANS DB, ADSAY V (2020) Variant anatomy of the biliary system as a cause of pancreatic and peri-ampullary cancers. *HPB (Oxford)*, 22(12): 1675-1685.

PINA LN, SAMOILOVICH F, URRUTIA S, RODRÍGUEZ A, ALLE L, FERRERES AR (2015) Surgical considerations of the cystic duct and Heister valves. *Surg J (N Y)*, 1(1): e23-e27.

RENZULLI M, BRANDI N, BROCCHI S, BALACCHI C, LANZA C, PETTINARI I, STEFANINI B, CARRAFIELLO G, PISCAGLIA F, GOLFIERI R, MARASCO G (2023) Association between anatomic variations of extrahepatic and intrahepatic bile ducts: Do look up!. *J Anat*, 242(4): 683-694.

RENZULLI M, BROCCHI S, MARASCO G, SPINELLI D, BALACCHI C, BARAKAT M, PETTINARI I, GOLFIERI R (2021) A new quantitative classification of the extrahepatic biliary tract related to cystic duct implantation. *J Gastrointest Surg*, 25(9): 2268-2279.

RENZULLI M, CARETTI D, PETTINARI I, BISELLI M, BROCCHI S, SERGENTI A, BRANDI N, GOLFIERI R (2022) Optimization of pineapple juice amount used as a negative oral contrast agent in magnetic resonance cholangiopancreatography. *Sci Rep*, 12(1): 531.

ROSKAMS T, DESMET V (2008) Embryology of extra- and intrahepatic bile ducts, the ductal plate. *Anat Rec (Hoboken)*, 291(6): 628-635.

SANGAMESWARAN K (2021) Variations of cystic duct and its clinical significance. *Int J Anat Res*, 9(4): 8120-8126.

SARAWAGI R, SUNDAR S, GUPTA SK, RAGHUWANSHI S (2016) Anatomical variations of cystic ducts in magnetic resonance cholangiopancreatography and clinical implications. *Radiol Res Pract*, 2016: 3021484.

SHAW MJ, DORSHER PJ, VENNES JA (1993) Cystic duct anatomy: an endoscopic perspective. *Am J Gastroenterol*, 88(12): 2102-2106.

TAN CE, MOSCOSO GJ (1994) The developing human biliary system at the porta hepatis level between 29 days and 8 weeks of gestation a way to understanding biliary atresia. Part 1. *Pathol Int*, 44(8): 587-599.

TAOUREL P, BRET PM, REINHOLD C, BARKUN AN, ATRI M (1996) Anatomic variants of the biliary tree: diagnosis with MR cholangiopancreatography. *Radiology*, 199(2): 521-527.

TSITOURIDIS I, LAZARAKI G, PAPASTERGIOU C, PAGALOS E, GERMANIDIS G (2007) Low conjunction of the cystic duct with the common bile duct: does it correlate with the formation of common bile duct stones? *Surg Endosc*, 21(1): 48-52.

TURNER MA, FULCHER AS (2001) The cystic duct: normal anatomy and disease processes. *Radiographics*, 21(1): 3-294.

UETSUJI S, OKUDA Y, KOMADA H, YAMAMURA M, KAMIYAMA Y (1993) Clinical evaluation of a low junction of the cystic duct. *Scand J Gastroenterol*, 28(1): 85-88.

XU YB, BAI YL, MIN ZG, QIN SY (2013) Magnetic resonance cholangiography in assessing biliary anatomy in living donors a meta-analysis. *World J Gastroenterol*, 19(45): 8427-8434.

ZONG Y, STANGER BZ (2011) Molecular mechanisms of bile duct development. *Int J Biochem Cell Biol*, 43(2): 257-264.