



## Case report

# Congenital absence of the cystic duct: Case report of a rare anomaly and review of the literature

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## ABSTRACT

**Introduction and importance:** Congenital absence of the cystic duct is one of the rare types of anomalies associated with the extrahepatic biliary tract (EHBT). It is often an incidental finding intraoperatively leading to significant implications during the perioperative period.

**Case presentation:** A 25-year-old lady was admitted for an elective laparoscopic cholecystectomy indicated for recurrent symptoms of right upper quadrant pain with evidence of cholelithiasis on ultrasound. During laparoscopy, the cystic duct could not be identified. After retrieval of the gallbladder, a blind ending orifice resembling an obliterated cystic duct was discovered.

**Clinical discussion:** Absence of the cystic duct can result from a congenital or an acquired process. In both cases, they are difficult to diagnose pre-operatively even though magnetic resonance cholangiopancreatography (MRCP) has shown great potential in delineating the EHBT. It confers an increased risk of injury to the surrounding biliary tract during cholecystectomy. Therefore, the surgical approach depends on the surgeon's operative competency and knowledge related to EHBT anomalies.

**Conclusion:** Definitive treatment for patients with symptomatic absent cystic duct is an open cholecystectomy, given its increased likelihood of iatrogenic morbidity. Nonetheless, it is important to highlight that laparoscopic cholecystectomy may be performed if the surgeon carries sufficient skills.

## 1. Introduction

The extrahepatic biliary tract is a common site for anatomical variations which has been reported to be as high as 47 % [1–3]. Previous studies such as those conducted by Kullman et al. [4] as well as Hasan et al. [5], have demonstrated anatomical variations of 19 % and 15 % respectively. These are significant results especially in consideration of the fact that cholecystectomy is the most common digestive tract surgery performed globally [6].

The focus of the case report revolves around the congenital absence of the cystic duct. This is a rare anomaly of the EHBT which was first described by Walton in the year 1925 [3,7]. Thereafter, only twelve cases have been documented in the literature [4]. It has significant implications intraoperatively and as such, during the postoperative recovery period as well [1–3]. Herein, we report a case of an absent cystic duct in a young patient with a history of recurrent right upper quadrant

pain. This case report has been reported in line with the SCARE criteria [8].

## 2. Case presentation

A 25-year-old female patient admitted as an elective case for laparoscopic cholecystectomy indicated for symptomatic cholelithiasis. The patient had a five-year history of recurrent presentation to the emergency department complaining of right upper quadrant pain associated with nausea and vomiting. Otherwise, she had no other past medical or surgical history and no hospital admissions prior. Laboratory investigations were insignificant as shown in Table 1. Ultrasound of the abdomen and pelvis showed a distended gallbladder, with a thickened wall and harbouring three stones (largest measured 2 cm). The common bile duct (CBD) was normal in diameter and no stones were visualised within it. Furthermore, the ultrasound showed a normal finding of the

**Abbreviations:** EHBT, extrahepatic biliary tract; CBD, common bile duct; MRCP, magnetic resonance cholangiopancreatography; ERCP, endoscopic retrograde cholangiopancreatography.

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**Table 1**  
Biochemical profile.

Test name	Unit	Test name	Unit
White blood cells	7.0	Albumin	44.2 g/L
Red blood cells	4.34	Alanine transaminase	10 IU/L
		Aspartate transaminase	12 IU/L
Hemoglobin	12	Alk. Phos	62 IU/L
Hematocrit	0.346	Direct Bil	2.8 umol/L
Mean corpuscular volume	79.6	Total bilirubin	6.3 umol/L
Mean corpuscular hemoglobin	348	Indirect bilirubin	4 umol/L
Mean corpuscular hemoglobin concentration	14.1	Urea	3.5 mmol/L
Red blood cell distribution width	283	Creatinine	56 umol/L
Platelet	283	Na (sodium)	136 mmol/L
		K (potassium)	4.1 mmol/L
		CL (chloride)	102 mmol/L

intrahepatic biliary ducts.

During the laparoscopic cholecystectomy, while trying to achieve the critical view of safety and identify the calot's triangle, the cystic duct could not be visualised. The cystic artery was identified, clipped and transected. The gallbladder was dissected fully from the liver bed and retrieved. There was no tract between the gallbladder and extrahepatic biliary tract even after meticulous dissection of the gallbladder from the liver bed. A drain was secured in the right subcostal region and closure of the surgical sites with primary repair was accomplished. The gallbladder was then examined which revealed three stones within its lumen (Fig. 1). However, a blind ending orifice was found within the specimen that might have resembled an obliterated cystic duct (Fig. 2). On the first postoperative day, the drain was found to contain minimal serous

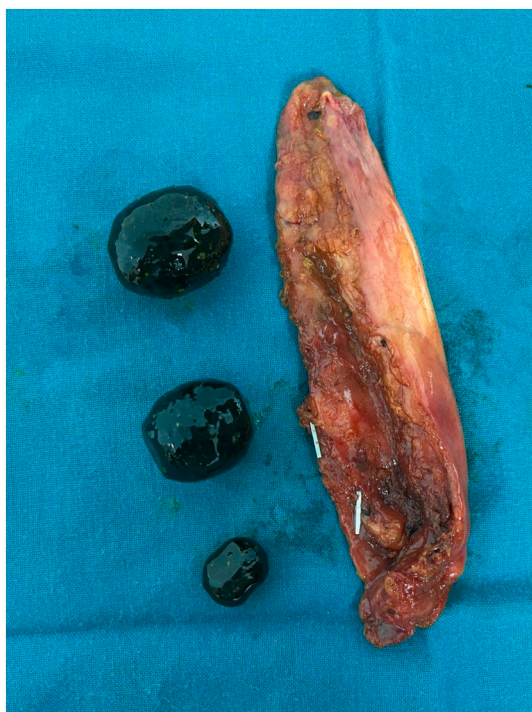


Fig. 1. The Gallbladder with its stones.



Fig. 2. The Gallbladder flipped inside out showing the blind ending (arrow).

discharge. The drain was removed and the patient was later discharged following an uneventful recovery. During her subsequent follow-up visits in the outpatient clinic at 2 weeks and 6 weeks post-operative, there were no issues concerning a bile leak or other complications.

### 3. Discussion

First of all, the cystic duct connects the gallbladder to the biliary tract and the site of its insertion marks the division between the common hepatic and common bile ducts [9]. It is approximately 2-4 cm in length and has a diameter of 1-5 mm [9]. There are concentric folds within the ducts known as the spiral valves of heister [9].

To understand the congenital anomalies associated with the biliary tract, one must reflect on the complex embryology of it. The first anlage of the biliary system arises from the hepatic diverticulum which develops during the fourth week [10,11]. This gives rise to the pars hepaticus which later forms the liver and intrahepatic biliary ducts [10,11]. It also gives rise to the pars cystica which forms the gallbladder, cystic duct and CBD [10,11]. Failure at any stage of this development process could result in the agenesis of the gallbladder and its duct. However, an absent cystic duct could also be an acquired pathology through recurrent inflammation and fibrosis. This is often attributed to gallstone impaction. According to the literature, one can differentiate between the two mechanisms through presentation, radiology and histopathology [6,12].

In a congenitally absent cystic duct, the patient presents with jaundice or acute pancreatitis with radiological findings of anatomical anomalies associated with non-inflammatory changes [6,12]. Histopathology would confirm the absence of the cystic duct. However, in the acquired process, the patient presents with biliary colic or acute cholecystitis [6,12]. There would be radiological findings of an inflamed gallbladder and an impacted stone at the neck [6,12]. Histopathology in this case would identify a remnant cystic duct with all its three layers [6,12]. To reflect on our patient's case, she presented with recurrent history of biliary colic, ultrasound showed no evidence of acute inflammation, and her histopathology was evident for chronic changes. One can conclude that there is validity with the literature review to some extent; nonetheless, there remains a significant lack of clear

understanding associated with this anomaly.

Congenital anatomic anomalies of the cystic duct often relate to the level and site of its insertion into the EHBT [13]. Level of insertion often occurs in the middle one-third of the EHBT in 75 % of individuals [13]. However, anatomic anomalies may exist where the cystic duct is found to insert in the distal one-third of the EHBT (10 %) and less commonly in the proximal one-third [13]. If it inserts distally, then it may run parallel with the common hepatic duct before fusion and may share a common sheath [13]. Site of its insertion is typically in the right lateral position of the EHBT, though variations may result in it inserting at the anterior or posterior site [13]. In some cases, the cystic duct may drain directly into the right hepatic duct, left hepatic duct or the ampulla of Vater [13]. Other unusual anomalies, documented in the past, include double cystic ducts and dual absence of both the gallbladder and its duct [13]. Finally, an absent cystic duct is also a rare occurrence with the gallbladder often emptying via a different pathway [2,3,13]. These anatomical anomalies can increase the risk of biliary injury; therefore, it is a surgeon's

responsibility to be familiar with the common anomalies of the biliary tree [2,3,13]. These are illustrated clearly in Fig. 3.

In 2013, Patil et al. conducted a literature review and gathered a total of twenty cases of cystic duct anomalies of which eleven were congenital [6]. In 2019, Kirov published the 12th congenital case [12]. As a result, it can be summarised that thus far, there are thirteen reported congenitally absent cystic duct cases including the case presented here. The average age amongst the thirteen cases is forty-nine with a greater incidence in females than males. As for the presentation, eleven of the thirteen cases had a history of recurrent attacks with a duration ranging from six months to twenty-five years. Complications such as jaundice and cholecystitis were also reported in seven of them.

A thorough literature review was conducted in order to aid the establishment of diagnostic approaches and preferred surgical management. The surgical approach utilized in all the cases prior to 2019 was a traditional antegrade or retrograde open cholecystectomy [6]. Intraoperative findings of the previous cases varied with some showing





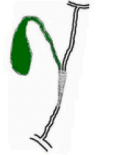


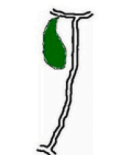

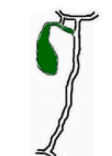


		
Lateral angular insertion (75%)	Anterior spiral	Posterior spiral
		
Low insertion (10%)	Parallel with CBD with common sheath (14 - 23%)	Insertion into ampulla of vater
		
High insertion	Insertion into right hepatic duct (0.006 - 0.01%)	Insertion into left hepatic duct
		
Cystohepatic duct (1 - 2%)	Double cystic duct	Absent cystic duct

Fig. 3. Anomalies of the cystic duct [6].

opening of the common bile duct and/or the common hepatic duct into the gallbladder [6]. In others, there was direct attachment of the gallbladder to the common bile duct [6]. Intraoperative complications were seen in three of the open cholecystectomies where the CBD was accidentally ligated and divided [6]. The case presented by Kirov and here, are the first two to be reported in the literature to have performed a laparoscopic cholecystectomy [12]. A great emphasis should be placed on the fact that there were no complications encountered during both laparoscopic cholecystectomies. On the contrary, the patients benefited from a faster recovery time and a shorter hospital stay.

As mentioned above, the absence of the cystic duct increases the risk of iatrogenic bile injury which may perpetuate to acute and chronic sequelae postoperatively. Therefore, it is worthwhile reviewing the imaging modalities that could be used to reach the diagnosis prior to surgery. According to the Radiological Society of North America, the cystic duct is not normally visualised on ultrasound [13]. The diagnostic method of choice for biliary tree anomalies is the use of MRCP or Endoscopic retrograde cholangiopancreatography (ERCP) [12–14]. ERCP is an invasive procedure and is largely reserved for patients requiring therapeutic interventions [13,14]. MRCP is the non-invasive imaging modality of choice, which is used to delineate the biliary tree anatomy [12–14]. Nonetheless, it is not a routine investigation prior to cholecystectomy due to the cost effect [15]. Therefore, a high index of suspicion is needed regarding the absence of the cystic duct for MRCP to be considered.

A greater focus should be attributed to the management of an absent cystic duct given that the majority of the cases are diagnosed intraoperatively [1–3]. Since the 1990's, the laparoscopic approach for cholecystectomies has widely replaced the open technique [16]. Therefore, most absent cystic duct anomalies will be incidentally discovered during laparoscopy. In this case, the conversion to an open cholecystectomy should be performed particularly if the anatomy is ambiguous [6,12]. However, if the surgeon has the knowledge and sufficient surgical skills, laparoscopic resection can be attempted [6,12]. Intraoperative cholangiography can be utilized to demonstrate the biliary anatomy and guide the surgery [6,12]. Ultimately, a clear assessment of the common biliary tree anomalies should be explored prior to ductal clipping. This additional precocious step could significantly decrease the risk of biliary tree injury and morbidity.

#### 4. Conclusion

Congenital absence of the cystic duct is a rare extrahepatic bile duct anomaly. The case reported here, is the 13th presented thus far in the literature. Such an anomaly arises from the pathological development of the pars cystica. The literature review, looking at all previous twelve cases, revealed that it seems to occur more often in females with a common presentation of recurrent right upper quadrant pain. It can also be associated with complications like jaundice and pancreatitis. It is often diagnosed intraoperatively even though MRCP has demonstrated great efficacy in the visualization of the biliary tract. The definitive treatment for patients presenting symptomatically is an open cholecystectomy. However, given that laparoscopic cholecystectomy has widely replaced open procedures, a surgeon with sufficient skills and knowledge may attempt a laparoscopic approach.

Documentation of such cases is highly encouraged in order to overcome the current limitations of resources available within the literature.

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#### Ethical approval

Not applicable.

#### Consent

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#### Author contribution

Reem Boushehry: literature review, writing, editing, manuscript drafting.

Fatemah Husain: writing, editing.

Athary Saleem: writing, editing.

Mohammed Alshamali: assisted in surgery, literature review, paper and picture editing, critical review.

Fahad Alhammadi: editing.

Khaleel Mohammad: performed the surgery, critical review, supervision, final approval.

#### Guarantor

Reem Boushehry.

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