

# Importance of Spontaneous Choledochoduodenal Fistulas Detected During ERCP Procedure

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**Abstract** Choledochoduodenal fistula (CDF) is an abnormal passage between the choledochus and duodenum. The most common causes of CDF are cholelithiasis, duodenal ulcer, and tumors. There are mainly two types of fistulas depending on the location. Type 1 is usually present on the longitudinal fold just close to the papilla. Type 2 is present at the duodenal mucosa adjacent to the longitudinal fold and probably caused by larger stones, duodenal ulcer penetration, impacted cystic duct stones, and as a complication of laparoscopic cholecystectomy. In this study, we investigate the characteristics of our patients those were diagnosed with CDF. This is a descriptive study. We retrospectively obtained the data of 21 patients with spontaneous CDF out of 2430 endoscopic retrograde cholangiopancreatography (ERCP) patients between 2000 and 2014. We analyzed the laboratory results, demographic and etiological features, major clinical presentations, diagnostic methods, and treatment modalities of the patients. The mean age of the 21 patients was  $66.6 \pm 2.2$  years and a female to male ratio was 12:9. In ten patients, interventional procedures were performed via fistulotomy, not through the papilla. The eventual diagnosis was tumor in five patients and stone or sludge in bile ducts in 14 patients. In the remaining two

patients, no reason was found as a cause of CDF. Whipple operation was performed in one patient and stents were placed in three patients for malignancy. Among the 14 patients with sludge or stone in bile ducts, ERCP has been therapeutic in ten. One of the remaining patients has been operated for proximal fistula and underwent choledochus exploration and repair of fistula over a T-tube. In the second patient, stone extraction and T-tube drainage were performed. In patients who had bile duct obstruction and got over of jaundice afterwards, one of the most important reasons of this recovery is the development of spontaneous choledochoduodenal fistula. Even if it is very rare, malignancy can be observed in this area. Therefore, it is extremely important to evaluate the papillary area with ERCP and to conduct biopsy; this will make early diagnosis possible in many patients. In these patients, ERCP can both be diagnostic and therapeutic.

**Keywords** Fistula · Choledochoduodenal · ERCP · Stent · Papillotomy

## INTRODUCTION

Internal biliary fistula is an abnormal communication between the biliary tract and adjacent organs [1]. The most frequent site is between the gallbladder and duodenum [2, 3]. Choledochoduodenal fistula (CDF) is an abnormal passage between the choledochus and duodenum. The most common causes of CDF are cholelithiasis, duodenal ulcer, and tumors [2–4]. There are mainly two types of fistulas depending on the location. Type 1 is usually present on the longitudinal fold just close to the papilla also named as parapapillary CDF. It is usually small and arose from an intramural portion of the common bile duct. It is reported that almost all parapapillary fistulas are caused by spontaneous extrusion of the stones in

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the choledochus [2–5]. It is formed by a stone small enough to enter the intramural portion of the common bile duct and also impacted the portion of the common bile duct. The rise in intraductal pressure causes ulceration and eventually leads to expulsion of the stone as formation of the fistula [6]. The passage of bougies or dilators into the duodenum during exploration of the common bile duct may also cause parapapillary CDF. That type of fistula is usually located distally and connected to the duodenum within 2 cm of the distal common bile duct. Type 2 is present at the duodenal mucosa adjacent to the longitudinal fold and probably caused by larger stones, duodenal ulcer penetration, impacted cystic duct stones, and as a complication of laparoscopic cholecystectomy. The fistula in type 2 is large enough for larger stones to pass in comparison to that in type 1. It is also located more proximally and drained from elsewhere of the biliary system (2 cm above the junction of the common bile duct to the papilla) into the duodenum. The cumulative frequency of both fistulas is 2.53% in endemic areas. It is three to four times higher in non-endemic areas. The other most observed causes of CDF are listed in Table 1 [7]. In this study, we investigate the characteristics of our patients that were diagnosed with CDF.

## Methods

We retrospectively obtained the data of 21 patients with spontaneous CDF out of 2430 endoscopic retrograde cholangiopancreatography (ERCP) patients at two referring hospitals. All procedures were performed by the same surgeon. We analyzed the laboratory results, demographic and etiological features, major clinical presentations, diagnostic methods, and treatment modalities of the patients. Duodenoscopy and ERCP procedures have been performed as described before in the literature [8]. The patients were placed in left-lateral position and a duodenal fiberscope was introduced into the duodenum by setting sun maneuver. Cannulation and visualization of the bile duct were performed with the patient in mostly the left lateral rarely the prone position. After the cannulation, a biliary system was visualized

by injecting the contrast material through the sphincterotome and the procedure needed was performed after that (Figs. 1, 2, 3, and 4).

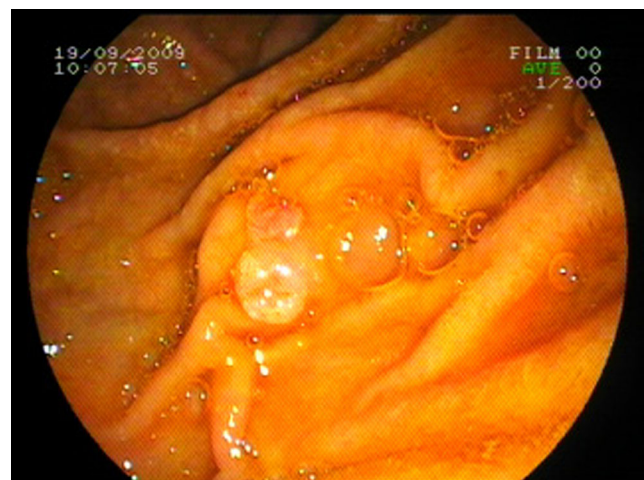
## Results

The mean age of the 18 patients was  $66.6 \pm 2.2$  years, and a female to male ratio was 12:9. Although the patients were presented with choledochoduodenal fistula, the diameter of the ducts was  $19.2 \pm 0.2$  mm. So the occurrence of the fistula did not relieve the enlargement of the ductus. The orifices of the CDFs were at 12 o'clock position in 11 patients, 11 o'clock position in 5 patients, and 3 o'clock position in 1 patient. In the last four patients, orifices were in atypical localization because of tumor. Leukocyte of the patients were slightly increased as  $11.5 \pm 1.3$  K/uL (4.0–10.0 K/uL), and liver function enzymes were compatible with obstructive jaundice as two to three times elevation [aspartate aminotransferase (AST):  $115.3 \pm 26.3$  U/L (0–34 U/L); alanine aminotransferase (ALT):  $139.5 \pm 28.9$  U/L (0–49 U/L)]. The enzymes indicating the cholestasis were also increased as follows: gamma-glutamyl transferase (GGT):  $440.2 \pm 37.3$  U/L (11–49 U/L) and alkaline phosphatase (AP):  $735.8 \pm 120.4$  U/L (50–136 U/L). The patients were severely icteric with bilirubin values of  $6.2 \pm 2.1$  mg/dL (0.3–1.2 mg/dL) and direct component predominance. Mean cancer antigen (CA) 19–9 values of the patients were also two to three times higher than the normal range as  $150.3 \pm 58.2$  U/mL (0–27 U/mL). In ten patients, interventional procedures were performed via fistulotomy, not through the papilla. The eventual diagnosis was tumor in five patients and stone or sludge in bile ducts in 14 patients. In the remaining two patients, no reason was found as a cause of CDF. Whipple operation was performed in one patient for periampullary tumor, and stents were placed in three patients (two SEMSs—self expandable metallic

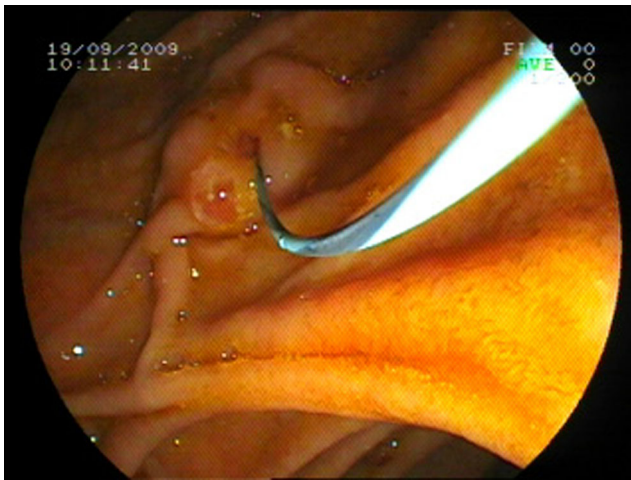
**Table 1** The causes of choledochoduodenal fistula

The causes of choledochoduodenal fistula

Cholelithiasis
Choledochostomy
Periampullar tumor invasion
Pancreatitis
Open or laparoscopic cholecystectomy
Perforated duodenal ulcer
Papillitis



**Fig. 1** ERCP showing the CDF orifice above the papilla

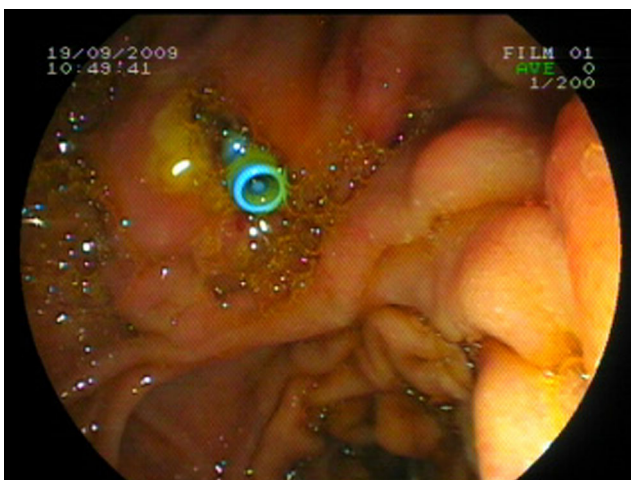


**Fig. 2** Cannulation of the fistula orifice

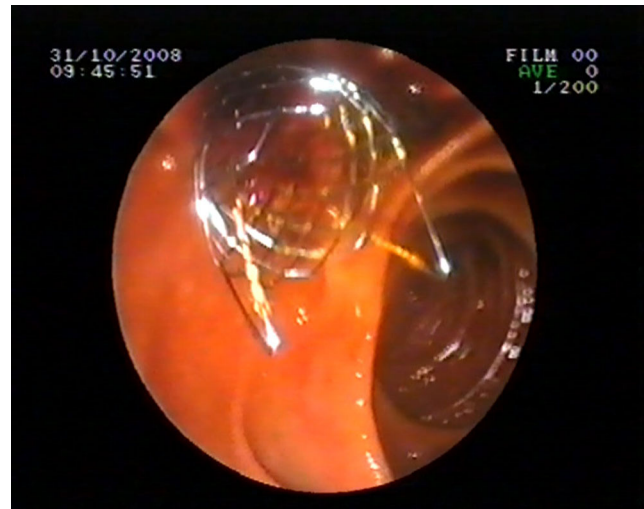
stents—for periampullary tumor through a fistula orifice and one plastic stent for a Klatskin tumor found incidentally in a patient with CDF). Among the 14 patients with sludge or stone in bile ducts, ERCP has been therapeutic in 12. One of the remaining patients has been operated for proximal fistula and underwent choledochus exploration and repair of fistula over a T-tube. This patient had been undergone cholecystectomy 6 years ago. In the second patient, stone extraction and t-tube drainage were performed.

## Discussion

Although the clinical presentation of the CDF varies depending on the location, most of the patients remain asymptomatic. However, in symptomatic cases, the cardinal presentation mimics those of cholelithiasis and choledocholithiasis who generally had right upper quadrant pain, fever, and jaundice. In our series, most of the patients presented with right upper quadrant pain, fever,



**Fig. 3** A plastic stent placed through CDF via ERCP



**Fig. 4** An SEMS placed through CDF via ERCP

and jaundice which were defined as cholangitis. ERCP findings were also compatible with cholangitis in those patients. Periampullary choledochoduodenal fistula may form a small blind cesspool at the common bile duct distal to the fistula similar to that observed in postoperative biliary sump syndrome. This presentation is named as obstructive cholangitis. But it is not clearly known that whether these symptoms are due to fistula or retained/recurrent biliary tract stones which can be dealt with endoscopic extraction successfully [6]. Moreover, the existence of fistula may contribute to the formation of bile duct stones due to infection through fistula. Although the diameter of the choledochus of our patients was nearly 20 mm, eight of them were found to have stones in the bile duct. The drainage of fistulas may serve as a compensatory mechanism for relieving the obstruction. The size of the fistula seemed to be the determining factor in diminishing the degree of jaundice. This condition was presented as lower bilirubin levels in proximal fistulas since they have larger diameter and better drainage than distal ones [7]. But the bilirubin levels of our patients were severely high as  $6.2 \pm 2.1$  mg/dL (normal level 0.3–1.2 mg/dL).

Periampullary CDF may be another mechanism that causes fluctuation of jaundice in ampullary carcinoma that can be observed rarely. In such a condition, obstructive cholangitis as well as obstructive jaundice may be evident. Therefore, when ampullary carcinoma produces obstructive jaundice and cholangitis, the presence of periampullary CDF should be taken into consideration and evaluation with ERCP is helpful and necessary in the diagnosis of tumor. But rarely if there are multiple parapapillary fistulas, jaundice may not be observed [2]. In our patients, if we consider the CA 19–9 levels, they were found to be increased cumulatively as  $150.3 \pm 58.2$  U/mL (normal level 0–27 U/mL). It appears that simple duodenobiliary reflux is asymptomatic as long as the refluxed material and bile can drain freely from the fistula [6]. But in selected cases, untreated fistula may be present with biliary sump syndrome presenting with cholangitis,

pancreatitis, abdominal pain, and fever. The large diameter of type 2 fistula seems to offer a route of such clinic.

The fistula itself may lead to recurrent cholangitis and/or pancreatitis by the same mechanism as that of biliary sump syndrome. Reflux cholangitis sometimes may occur in patients with untreated type 2 fistulas without recurrent stones, but it may be treated quickly with conservative treatment. Chronic cholangitis due to reflux of duodenal contents with activated pancreatic juice and bacterial flora into the bile ducts may be a causative factor even for the development of bile duct cancer [9]. Repeated reflux of pancreatic juice may be a stimulating factor in premalignant and malignant changes of biliary epithelium and eventually results in development of biliary tree and gallbladder carcinoma [1, 5]. It has been well demonstrated before that biliary-enteric anastomoses induce reflux of intestinal juice into the biliary tract and play a role in biliary carcinogenesis. As a result, high incidences of secondary biliary carcinoma have been observed in patients who have undergone biliary reconstruction [10, 11].

The diagnosis of the patient with CDF can be made by conventional radiological methods and a duodenal fibroscope. It causes air in the bile duct on plain X-ray [5, 12]. The direct observation as well as the exit of the cannula from the papilla through fistula confirms the diagnosis. Successful cholangiography performed through the papilla as well as the fistula orifice also confirms the diagnosis during the ERCP studies [4]. In ERCP-based series, the frequency of CDF near the papilla of Vater is much greater in patients with hepatobiliary and pancreatic diseases [3]. Distal common bile duct stones located in the space left between the fistulous orifice and the orifice of the papilla may occur due to stasis of the lithogenic bile [13], so the patients usually have increased levels of alkaline phosphatase (AP) and gamma glutamyl transferase (GGT).

There are several conflicting reports about the treatment of CDF. Proximal fistulas should be treated surgically, whereas endoscopic therapy should be tried first for distal ones [7]. The management of distal CDF is connecting the natural orifice with the fistula or enlarging the fistula in a cephalad direction [14]. After therapeutic endoscopic sphincterotomy, the flow of air and duodenal contents through fistula and also cholangitis are not observed frequently. Currently, most patients with peripapillary CDF undergo endoscopic sphincterotomy (ES) for the treatment of accompanying bile duct stones rather than operative procedures such as transduodenal papillotomy or papilloplasty. However, the most appropriate treatment for patients with a fistula but without stones is unknown [9]. The incidence of stone recurrence is not significantly different between patients who underwent ES and those remained untreated [9]. Uncomplicated peripapillary CDF probably does not require treatment [15]. ES has been considered standard for the treatment of fistula if bile duct stone is present. Some investigators have described the effectiveness of enlarging the fistula in a cephalad direction through fistula orifice with

the ES technique. Fistulotomy is necessary and sufficient, but there is no need for papillotomy. In difficult cases, a needle-knife papillotomy cutting from the fistula orifice to the natural ampulla may be needed [16]. However, some authors claim that since the majority of these defects may be the complication of cholangitis and bile duct stones, surgical management should be considered even if the patient has no significant clinical symptom [17]. As a conclusion, our patients with choledochoduodenal fistula presented with cholangitis symptoms, stones in bile ductus, and increased levels of liver enzymes and bilirubin values as opposed to the reports published before. So we believe that the patients with choledochoduodenal fistula should be initially managed with ERCP before any kind of surgical procedures, since they may have recurrent bile duct stones or malignant tumors and in these kind of patients, ERCP can both be diagnostic and therapeutic.

#### Compliance with Ethical Standards

**Conflict of Interest** The authors declare that they have no conflict of interest.

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