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Chapter

# Role of Endoscopic Retrograde Cholangiopancreatography in Benign Biliary Diseases

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## Abstract

Endoscopic retrograde cholangiopancreatography (ERCP) is a combination of endoscopy and X-ray technique, which was introduced as a diagnostic tool but with the advancement in the technology such as balloon dilatation or stent placement in combination with ERCP has transformed the latter into therapeutic accessory for multiple biliary diseases. It can also be used as an adjunct tool to increase the success rate of therapy. This diversified application of ERCP emphasizes the importance of this procedure for patients with biliary diseases despite the certain post-interventional complications. The scope of ERCP procedure is continuously increasing in the detection of anatomical or physiological abnormalities. ERCP plays an important role in conditions with biliary obstruction or biliary leaks, which may be due to primary or secondary causes. Biliary stents can be placed in combination with ERCP, which can assist in achieving therapeutic goals in patients with biliary strictures or clearance of biliary sludge.

**Keywords:** endoscopic retrograde cholangiopancreatography, ERCP, benign biliary disease, choledocholithiasis, cholangitis

## 1. Introduction

Benign biliary diseases are diversified range of disorders, which may be congenital or acquired that can impact hepatic bile ducts and liver involvement. Incident diagnosis is very common in benign biliary diseases as the symptoms can vary from clinically asymptomatic to cholestatic symptoms with acute or chronic clinical path resulting in malignancy [1]. Patient workup is very important in determining the etiology of the disease, suggestive symptoms play a key role such as abdominal pain with fever can be due to choledocholithiasis, prior history of hepato-biliary surgery leading to biliary leak, autoimmune diseases, inflammatory strictures, and family history hereditary/ congenital disorder [1].

Endoscopic retrograde cholangiopancreatography (ERCP), which uses combination of endoscopy and X-rays, was first introduced as a safe technique for diagnostic purpose of pancreaticobiliary disease. With the addition of endoscopic sphincterotomy technique, ERCP developed into therapeutic tool [2]. It is now considered to be gold standard for diagnosis of biliary-related diseases but also more reserved for

Benign Biliary diseases	
Cholelithiasis or gallstone disease	Choledocholithiasis
Post-surgical biliary complications	Bile leaks
	Biliary strictures
Stictures	Primary sclerosing cholangitis
	IgG4-related sclerosing cholangitis
	Ischemic cholangitis
	Inflammatory strictures
Developmental anomalies	Caroli's disease
	Choledochal cysts
	Sphincter of Oddi dysfunction
Others	Mirizzi Syndrome
	Parasites

**Table 1.**  
*Types of benign biliary disease.*

therapeutic purpose owing to its more invasive nature when related to the alternative diagnostic tools [3]. Multiple imaging techniques are used in diagnosis for biliary disease such as intraoperative cholangiography, abdominal ultrasonography (US), computerized tomography (CT), magnetic resonance cholangiopancreatography (MRCP), and endoscopic ultrasound (EUS). They also play key role in determining the patient selection for ERCP to be considered as therapeutic intervention [4], but still the role of ERCP in differential diagnosis of multiple disease cannot be ruled out. ERCP can also be used in combination with other techniques such as cholangiopancreatography where it applies as an adjunct for evaluation and management of biliary and pancreatic disease [5].

Endoscopists require extensive training to perform ERCP in order to increase the procedure success rate and also minimize as post-procedure complications such as bleeding, perforation, and pancreatitis which may lead to mortality and morbidity [6]. ERCP is precisely advantageous in the achievement of therapeutics in biliary obstructive patients due to choledocholithiasis, biliary leaks, or strictures where the success rates are found to be greater than 90% [7]. ERCP combined with bile duct stenting and/or biliary sphincterotomy is preferred when the diagnosis is bile leaks. Pre- and post-procedure antibiotic prophylaxis is recommended to all patients undergoing this procedure (Table 1).

## 2. Cholelithiasis or gallstone disease

Stone formation and presence in the biliary tree are referred to as cholelithiasis. This disease is one of the leading causes of hospital admissions worldwide and found to have prevalence rates of 3.2%–15.6% in Asia [8]. Stone location is the key to segmentation of this pathological condition as stones found in gall bladder and when the stones are located at extra hepatic bile ducts, it is referred to as choledocholithiasis, and intrahepatic stones presence causes hepatolithiasis. Stone formation is mainly due due to biliary stasis, which may be due to chemical imbalance of bile constituents or

impaired gall bladder emptying. There are several risk factors for cholelithiasis, both modifiable and non-modifiable. Obesity is one of the major risk factors for cholelithiasis while other risk factors include dyslipidemias and insulin resistance. Women are more prone to this pathological condition, whereas it has symmetrical relationship with the increase in age and is found to be asymptomatic in 80% of patients. Patients present with upper quadrant abdominal pain and positive for Murphy's sign. Abdominal ultrasound is found to be the most effective/noninvasive tool for diagnosis. The preferred treatment option for cholelithiasis is laproscopic cholecystectomy as an early intervention [9].

## **2.1 Choledocholithiasis**

Choledocholithiasis is referred to a condition when stones are present in common bile duct (CBD). Exact prevalence is unknown, but studies have reported that about 5%–20% patients diagnosed with cholelithiasis have stones in CBD [10]. Choledocholithiasis may be classified into primary or secondary types where secondary refers to stones passage to bile ducts, and primary is due to conditions predisposing to bile stasis, which may be due to cystic fibrosis or long-term total parenteral nutrition. Choledocholithiasis might be due to complication of cholecystectomy procedure, which can be detected about after 3 years of surgery. Recurrent choledocholithiasis can also occur post cholecystectomy, which might be due to various secondary causes that include rapid weight loss (bariatric surgery), sickle cell anemia, periampullary duodenal diverticulum, chronic cholangitis, and dilated CBD or CBD stricture. Clinical features that distinguish choledocholithiasis are long-lasting post-prandial right upper quadrant (RUQ) pain that may exceed 6 hours, which may radiate to epigastrium or even back. Extrahepatic cholestatic signs may also be present, e.g., dark urine, pale stool, pruritis, etc. Evaluation should be done in all patients with or without presenting symptoms. First line of diagnostic tools are liver functioning tests (LFTs) and abdominal ultrasound. ERCP is the preferred diagnostic and therapeutic tool for confirming the likelihood of choledocholithiasis. ERCP has been reported to be very sensitive and specific in detection of CBD stones with the success rates of more than 95%, smaller stones may still be missed [11]. Air bubbles into biliary ductal system can lead to altered or misdiagnosis of stones; hence, introduction of air bubbles and over-filling of ducts by contrasts injections should be avoided. Characteristic findings for choledocholithiasis smooth-walled, well-defined, intraluminal defects within CBD, which may or may not be dilated. Management of choledocholithiasis initially includes support therapy for patients with acute symptoms with further identification of complications and management. Definitive treatment for choledocholithiasis is removal of CBD stone or elective cholecystectomy [10, 11]. In patients with no cholangitis or biliary obstruction, it is recommended to delay the ERCP procedure to >48 hours as it does not reduce the mortality rate when compared with conservative treatment [10]. However, it is reported that in patients with concomitant cholangitis with biliary obstruction, early ERCP reduces the mortality as well as adverse events following the procedure [12, 13]. Major complication that is found post ERCP is pancreatitis, which can increase the chances of mortality, which limits the use of ERCP in particular patients. This occurrence of complication like pancreatitis can be lowered with generous use of intravenous hydration and nonsteroidal anti-inflammatory rectal suppositories. For removal of larger stones, sphincterotomy combined with balloon sphincteroplasty and mechanical lithotripter can be used with high success of larger and difficult stone removal [14]. Moreover,

where available, electrohydraulic lithotripsy may be used. However, if endoscopists failed to clear bile ducts, then plastic stent insertion is warranted for biliary drainage.

### **3. Post-surgical biliary complications**

As the technological advancements and increase in use of laparoscopic techniques with shortened recovery times, decreased size of abdomen incision, it has become one of the first choices for treatment of biliary-related diseases. Concomitant use of laproscopic interventions has arisen a new complication of bile duct injury (BDI), which prevails in 0.2–1.4% of patients undergoing laparoscopic cholecystectomy [15, 16] and is reported to cause complications such as cystic duct leakage, CBD leakage, or bile duct strictures. Majority of post-surgical biliary complications can be treated successfully in 75–90% of patients by endoscopic or radiological interventions [17, 18].

#### **3.1 Biliary leaks**

Leakage of biliary constituents into abdominal cavity due to hole in bile duct, which may be due to postoperative complication. Biliary leaks can occur within a week of surgery but may present with symptoms even after a month of surgery. The symptoms present in the patient with such condition are nausea, vomiting, RUQ pain, jaundice, anorexia, and fever [19]. A distinct collection of bile outside biliary tree also known as biloma is a distinguished presentation of biliary leak, which can be encapsulated due to inflammatory reaction and fibrosis. Several imaging modalities can be applied for diagnosis of post-surgical biliary leaks or bilomas with abdominal ultrasound being the initial imaging tool for quick and efficient follow-up for collection of biliary fluids [19]. Biloma can be present as ascitic fluid collection or well-confined loculated within particular boundaries, which is suggestive of an infection [20]. Computed tomography can also be used to detect bilomas and assess surrounding tissues where it can be used to study the further complications, e.g., biliary peritonitis. Source of biliary leakage can be identified by MRI and hepatobiliary cholescintigraphy [19]. Management of biliary leakage involves diversion and drainage of bilomas. Decompression of bile duct by sphincterotomy alone with or without endoscopic stent or nasobiliary drain placement. Stents are placed for 4–6 weeks, but larger duct injuries require longer duration of placement for healing of biliary leaks [21]. Biliary leaks can be segregated as low or high grade based on the extent of leakage after cholangiography, which can be identified during opacification. High-grade leaks are highly evident, whereas low grade requires complete filling of intrahepatic ducts to exhibit contrast extravasation. Location and grade of leak greatly influence the success rates of treatment with endoscopic procedure with range of 80–100% [22]. If ERCP and stenting fail, surgical correction is required.

#### **3.2 Biliary strictures**

Biliary strictures are a pathophysiological condition, which refers to constriction of intrahepatic or extrahepatic biliary ductal system. When this condition occurs, it hinders the normal flow of bile causing retention on bile and proximal dilatation, further causing biliary obstruction. Biliary strictures that are acquired are more common than congenital causes. Acquired biliary strictures are further classified into benign and malignant where 30% of biliary strictures are benign. During laparoscopic



cholecystectomy, bile duct injury is due to misidentification of biliary duct for cystic duct that causes injury and clip application, which leads to formation of biliary strictures later on. Bile duct injury due to laparoscopic cholecystectomy accounts for 0.7% of total incidence of biliary strictures [23]. Recognition of blood supply is of prime importance for therapeutic procedures [24]. Anastomotic biliary strictures are complications arising from orthotopic liver transplantation or Whipple procedure (incidence rate is found to be 4%) performed for pancreatic mass or tumor [24, 25]. Biliary strictures can also be due to infections such as tuberculosis. However, the most common etiology for biliary strictures is malignancies [26]. Underlying etiology has shown to have strong impact on the prognosis of condition with strictures arising due to malignant, and primary sclerosing cholangitis may have unfavorable prognosis, whereas chronic pancreatitis, trauma, radiation, or operative injury has a good prognosis. Strictures identified in an early stage respond better to endoscopic treatment, which involves serial placement of single or multiple plastic stents over a period of 1 year. Fully covered metal stents are available for benign strictures and can be removed later [27, 28]. Success rates for this procedure range from 74 to 90% but have a very high recurrence rate after the removal of stents, a more aggressive approach is found to have more consistent results. Late anastomotic strictures require long term and multiple therapies (1–2 years). Balloon dilation is found to be less effective than combination of balloon dilation and stent placement, which has response rates of 70–100%. Subsequently increment in the number of stents post ERCP is reported to be most effective therapeutic approach [29]. Post-surgical biliary-enteric anastomosis can be treated successfully with ERCP, which might be assisted with enteroscopy for stricture site access.

#### **4. Sclerosing cholangitis**

Sclerosing cholangitis is spectrum of pathological condition, which encompasses the inflammation of intrahepatic and/or extrahepatic bile ducts, which is progressive. Fibrosis, stricturing, and patchy inflammation are characteristic presentation of sclerosing cholangitis. The course of diseases varies greatly in involvement of bile ducts complicated by carcinoma even at early stages of disease or subtle occurrence of portal hypertension leading to cirrhosis and hepatic failure [30]. Sclerosing cholangitis can be differentiated into different types based on causative agents or factors ranging from unknown to known factors such as infections, pancreatic disorders, etc.

##### **4.1 Primary sclerosing cholangitis**

Primary sclerosing cholangitis (PSC) is the most common form of cholangitis. Exact etiology is unknown, but some of the literatures associate this with autoimmune conditions such as ulcerative colitis (UC). Inflammatory bowel disease accompanies PSC in 90% of patients, where 87% of patients have comorbidity with either Crohn's disease or ulcerative colitis. However, approximately 5% of UC and  $\leq 5\%$  of Crohn's disease develop PSC. Cholangitis is initially asymptomatic, which after progression shows signs of cholestasis and in the later stages may transform to cirrhosis. Males are more prone to this condition in comparison to females [31, 32]. Approach for diagnostics should be focused on the laboratory reports and radiological tests where persistent elevated cholestatic enzymes are sign of presence of PSC. Biliary obstruction is corrected with ERCP to clear stenosis. Multiple stenting might

be required for certain patients. ERCP is discouraged in the diagnosis of PSC due to its possibility of adverse events such as bacterial cholangitis, perforation of biliary tract, and pancreatitis. However, diagnostic use of ERCP in PSC has proven to be advantageous in certain conditions. It may facilitate the diagnosis of PSC, which is not determined by magnetic resonance cholangiopancreatography due to suboptimal imaging of intrahepatic bile ducts [33]. Secondly, it can also determine the prevailing stricture with high accuracy in patients with deteriorating conditions, which may present as persisting cholestasis, jaundice, or bacterial cholangitis. ERCP can also precisely establish existing cholangiocarcinoma, which can be misidentified for biliary stricture that possesses symptoms of biliary dilatation [33]. Strictures that are associated with PSC have positive prognosis with endoscopic intervention such as balloon dilation, which can be accompanied by stent placement. Since there are high chances of adverse reactions or complications, balloon dilatation is found to be sufficient, but the literature data are limited. Therefore, stent placement is preferred for dominant strictures, which are unmanageable by dilatation leading to increase in the chances of survival of patients with PSC.

#### **4.2 IgG4-related sclerosing cholangitis**

IgG4-related sclerosing cholangitis (IgG-SC) is a fibro-inflammatory disease with systemic involvement with classical findings for lesions and fibrosis in the biliary system. IgG-SC may have symptoms similar as PSC or pancreatic adenocarcinoma. Pathology of the condition varies from inflammatory stage to fibrosis to organ failure and even mortality. Epidemiological data for IgG-SC are very limited. IgG-SC is predominantly found in males usually affecting in fifth and sixth decade of life [34]. Epidemiological studies report chronic exposure to “blue collar work” to be associated with incidence of IgG-SC [35, 36]. Clinical history for chronic allergy with elevated levels of IgE directly correlates with the occurrence of IgG-SC. Clinical presentation for IgG-SC depends on the organ involved and the extent of disease. IgG-SC can affect any part of biliary tree. Obstructive jaundice, weight loss, and RUQ abdominal pain are associated with the presence of IgG-SC. Patients with IgG-SC can be spontaneously identified by abnormal liver functioning test and elevated markers for inflammation. Total IgG concentrations may or may not be elevated despite IgG-4 subclass concentrations being elevated [33]. ERCP alone as a diagnostic tool for IgG-SC has low level of accuracy to differentiate between IgG-SC and PSC or cholangiocarcinoma where cross-sectional imaging plays an important role in identification of IgG-SC. ERCP and biliary stenting help in symptomatic relief.

#### **4.3 Ischemic cholangitis**

Injury to any vessel or vessels supplying blood to biliary tract can causing impaired blood supply, which can be focal or extensive depending on the extent of injury. Formation of lesions in biliary system due to restricted blood flow is labelled as ischemic cholangitis (IC) [37]. Blood flow restriction that can cause IC can be at the level of major hepatic arteries or at microvascular plexus level. Damage to the vessels during liver transplantation or radiation therapy is among few of the common causes of IC. Hypercoagulative disorders can also cause thrombosis resulting in IC. The underlying factors related to ischemic biliary damage are arduous to identify as it has high mortality rate [38]. Lesions due to IC can be subdivided into extrahepatic and intrahepatic lesions or combination of both due to success rates of therapy. IC may

be asymptomatic in the initial stage with majority of IC showing signs post 6 months to a year of post-surgical intervention, with rarely secluded extrahepatic or intrahepatic involvement. Major presenting signs of IC are cholangitis and cholestasis, which are aggressively progressive leading to hepatocellular failure. Ischemic injury is prominent factor for recurrent signs of cholangitis. IC is not reversible, and management options are very limited. Uncomplicated IC is not associated with high risk of mortality but is likely to have high morbidity due to extensive invasive procedures of multiple dilatations and hospitalizations. Diffused IC requires ERCP with stricture dilatation and stent placement. Application of percutaneous transhepatic cholangiography-guided drainage if ERCP fails or hepaticojejunostomy may be required in conditions that are difficult to manage [39]. Endoscopic therapy is considered to be first line of IC strictures and is minimally invasive; it can also play an adjuvant role in bridging during liver retransplantation. Process of ERCP dwells removal of biliary sludge and casts. Since biliary strictures in IC are bilobar, diffused, and accompany high predilection for intrahepatic ducts, insertion of stent post balloon dilation is required [40]. Patients with IC may require stent replacement every 3–4 months [41].

#### 4.4 Recurrent pyogenic cholangitis

Biliary tree can be infested by parasitic organisms causing inflammatory disease leading to scarring of bile duct tissue ultimately followed by bile stasis, intrahepatic stones, stricture formation, and even portal hypertension. This condition is also known as pyogenic cholangitis, since it is associated with recurrent attacks; it is referred to as recurrent pyogenic cholangitis (RPC) [42, 43]. Hepatolithiasis can also cause RPC, which further leads to recurrent bacterial infections and biliary inflammation. It is also known as oriental cholangiohepatitis, exact etiology is unknown, but some literature reports association with *Ascaris lumbricoides* and *Clonorchis sinensis*. The involvement of parasites is unclear in the etiology of RPC, where data suggest debilitation of immunity allowing bacterial infestations, scarring, and fibrosis and ultimate stricture formation. Epidemiological data report RPC to be prevalent in Asian population, but it can also be found in American regions. Recently, the incidence rate of this disease is found to be in declining phase due to increment in the habitat standards and Westernization of diet. Therefore, demographic details play an important role in diagnosis of the disease. Common laboratory findings report elevated leucocyte count and levels of bilirubin. Patient suffering from RPC shows a much diversified range of symptoms, mild symptoms to fulminant abdominal sepsis. Radiological techniques are further used for confirmation of presence of RPC. Previously, direct cholangiography was considered to be first line, but ERCP has shown certain advantage over the former where extrahepatic strictures can be well identified using ERCP, and it permits better evaluation of peripheral ducts due to better spatial resolution. It provides both evaluation for diagnosis and therapeutic intervention in a single procedure. The aim of therapy is to achieve unobstructed flow of bile by eliminating calculi from biliary tree and prevention of further stone formation. In patients with extractable stones ERCP technique, which is reported to effective in 90% of the cases [42, 43]. Balloon angioplasty catheters can also be used for stricture dilatation, and plastic stents may be inserted to ensure integrity of decompressed duct. Fully covered metal stents can also be used because of their long term patency; however, because of benign nature of disease and high cost of metal stent, therefore, it is not recommended to use metal stents in this condition [44].



## **5. Developmental anomalies**

Pancreaticobiliary tree developmental anomalies may remain until later stages of adolescence or early adulthood. Unexplained signs and recurrent symptoms related to biliary tree issues such as abdominal pain, jaundice, nausea, and vomiting should trigger the presence of congenital anomaly and cholangiopancreatography should be warranted. Detection of developmental anomalies may facilitate in therapeutics and prevention of ductal injury. Developmental anomalies of biliary tree include Caroli's disease, choledochal cysts, and Von Meyenburg complexes.

### **5.1 Caroli's disease**

Caroli's disease (CD) is congenital anomaly of intrahepatic bile ducts accustomed by segmented cystic dilation of ducts. It is reported to develop from abnormal ductal plate malformation during developmental phase of biliary tree. It is theoretically explained to follow autosomal dominant inheritance in some families. CD can remain unnoticed during the first stages of life and can also remain the same for whole life. Dominant symptoms are progressive recurrent cholangitis, intrahepatic calculi, and abscesses and may also lead to sepsis. Liver transplantation is the definite treatment and required in some cases [45]. Males and females are equally prone to this condition with 80% of the cases being identified before the age of 30 [46, 47]. ERCP is reported to be very highly sensitive for diagnosis of CD perhaps some of the literature reports it to be of highest sensitivity [48]. Saccular dilatation is the distinguishing sign that confirms the presence of CD as the symptoms can be misidentified for PSC or RPC. Due to the distinguishing feature, identification of cystic lesions across the biliary tree is an important factor in diagnosis, which is accurately accomplished by ERCP. Positive evaluation of transient recovery from cholangitis by ERCP also gives an additional benefit of utilizing ERCP procedure in CD.

### **5.2 Choledocal cysts**

Choledocal cyst (CC) is one of the benign anomalies of congenital origin, which is associated with dilatation of intrahepatic or extrahepatic bile ducts. Approximately 1% of benign biliary diseases are CC [49]. It has high prevalence in Asian population with predominance in females. The incidence literature is very scarce with few studies reporting to be one in 13,000–2,000,000 live births. CC can be diagnosed in any part of life, but approximately 50% of the cases are diagnosed in the first decade of life. Many hypotheses have been proposed regarding origin of CC but the widely accepted is the association with abnormality in the arrangement of pancreaticobiliary ductal junction during development [49]. The pancreaticobiliary junction is situated near to the sphincter of Oddi, this common pathway causes retrograde flux of pancreatic juice into biliary tree causing inflammation, ectasia, and dilatation. Clinical presentation includes classic biliary symptoms in adults and with abdominal mass as distinguishing feature for CC in infants. But, this abdominal mass is physically palpable in approximately up to 60% of cases [50, 51]; therefore, diagnosis of CC cannot be ruled out in the absence. CC is segregated in different types according to the involvement of portion of biliary tree. The most common complication of CC is stone formation and malignancy. ERCP is considered to be gold standard for diagnosis of CC as it is found to be safe of patients of

all ages. ERCP can play therapeutic role in CC where biliary obstruction is found, whereas it is equally effective in giving clear picture of ductal anatomy for prior strategizing for definite surgical intervention. ERCP not only is effective in preoperative phase, but it can play important role in post-surgical phase in clearance of biliary sludge and monitoring of integrity of biliary tree [50, 52].

## 6. Others

### 6.1 Sphincter of Oddi dysfunction

Sphincter of Oddi (SD) is a muscular valve situated in the duodenum, which regulates the flow of pancreatic or hepatic contents into the small intestine. Prevention of accumulation of bile sludge and particulate matter is one of the distinguished functions, thus reducing the probability of inflammation. Failure to perform this function is known as sphincter of Oddi dysfunction (SOD). Prevalence of SOD is found to be 1.5% in general population, which increases to 23% in patients showing signs of biliary disease [53, 54]. SOD may occur in adult or pediatric population, but it is found to be more prevalent in middle-aged women [54]. SOD might relate to a prior cholecystectomy, which changes the dynamics of biliary system. Symptoms of SOD and gall bladder dysfunction cannot be distinguished; therefore, proper investigations are required to diagnose SOD. Classic biliary disease symptoms are also found in SOD, presence of which is the basis of classification of SOD. There are multiple diagnostic (invasive and noninvasive) methods that can be useful in identifying SOD, but gold standard is the use of Sphincter of Oddi manometry (SOM), which observes the motor activity of SD with the use of ERCP technique [55]. Despite technical hassle and expertise, SOM is widely accepted and used for diagnosis of SOD. SOM is indicated in patients with unexplained excruciating pancreatobiliary pain with or without abnormal liver enzymes. Some literature reports also suggest that SOM can also predict the outcome of sphincter ablation in the SOD patients [56]. The therapy for SOD is evolving with major aim to establish reflow of biliary or pancreatic content to intestine. Since there is very limited evidence of medical therapy, traditionally surgical therapy was preferred choice. However, use of less invasive laparoscopic techniques is preferred. Transduodenal biliary sphincteroplasty with pancreatic septoplasty is the most common surgical intervention but due to patient tolerance, cost of care, morbidity, and mortality, this approach is being minimized. At present, surgical intervention is only reserved for patients who have undergone through endoscopic procedure previously and symptoms have reemerged or if the endoscopic procedure is not feasible [56]. However, for pancreatic sphincter hypertension, surgical intervention is considered to be standard of care [57]. ERCP is termed as standard for type I and II SOD as it responds with positive etiology to ERCP [58]. Endoscopic sphincterotomy related to sphincter ablation is reported to have clinical improvement in about 90% of patients with SOD. Pancreatic sphincter hypertension is related to the failure of endoscopic interventions in patients as pancreatic sphincter pressure is unaltered even when biliary sphincter pressure is compressed [59]. Balloon dilatation and stent placements, which can become very common in GI strictures, are not currently known to be useful in SOD due to limited evidence [56]. Literature data are suggestive that ERCP along with manometric evaluations is the current standard for diagnosis and predictive of further therapy associated with SOD.

## **6.2 Mirizzi syndrome**

Mirizzi syndrome (MS) is a complication of long-term cholelithiasis, with the prevalence of 0.05–2.7% among high-risk patients with calculus of gallbladder [60, 61]. MS is the presence of gall stones in gall bladder or cystic ducts that causes extrinsic compression of common hepatic duct. Due to compression of ducts, the process of inflammation is initiated. Chronic inflammation leads gall bladder to shrink and partially fused with hepatic duct. Impaction of gall stone and shrunken gall bladder results in stricture and pressure necrosis of intervening wall, ultimately fistula formation [62, 63]. Clinical presentation for MS is features of obstructive jaundice and abdominal pain with or without any pathognomonic features on history and physical examination. Classification of MS is based on ERCP finding according to the positioning, presence of stone and fistula. Females are more prone to MS in comparison to males. This gender affinity toward females is associated with the higher incidence of gall stones in female gender [64]. Diagnosis in the earlier stages of MS is very significant as unidentified MS may result in biliary duct injuries as a consequence [65]. Currently, ERCP is gold standard for diagnosing MS as it also gives information regarding the cause and extent of biliary obstruction and the damage to the ducts. ERCP also distinguishes ductal abnormalities including presence of fistulas [66, 67]. Percutaneous transhepatic cholangiography (PTC) can also be used for diagnosis of MS, but it has less efficiency than ERCP, which can identify a low lying stone in cystic duct that is often missed by PTC. Furthermore, visualization of distal common biliary duct is hindered if there is obstruction at any level of common hepatic duct, whereas ERCP can also be used as therapeutic procedure for MS. Open surgical management is the standard of therapy in MS with positive results reported and decrease in overall mortality and morbidity [68]. Endoscopic procedures with therapeutic goal are reserved for patients who are poor candidate for surgery (e.g., elderly with multiple comorbidities) where stent placement can achieve the patency of common hepatic duct [69].

## **6.3 Biliary worms**

Parasitic infections are common in biliary tree, which are due to manifestations of nematodes and hermaphroditic trematodes. These parasitic infestations may cause diversified pathologies ranging from cholelithiasis to pancreatitis to liver abscess [70]. Presence of worms in stools and duodenal contents confirms the diagnosis of biliary infestation. ERCP is a very useful tool not only in diagnosis but also aims for removal of biliary worms. More than 80% of patients with biliary ascariasis are treated successfully by ERCP [71]. Treatment of ascariasis may or may not require sphincterotomy and balloon dilation. Both of which can be performed in a single session of ERCP setting. Children are more prone to parasitic infestations, and ERCP is also found to be safe and effective for this population as well.

## **7. Conclusion**

Endoscopic procedures are now preferred over surgical interventions due to less invasive in nature, cost of therapy, and patient compliance. ERCP plays an important role in diagnosis and therapeutics of multiple benign biliary diseases. Despite risk of post-procedural complications such as pancreatitis, bleeding, or perforation,

it is considered to be one of the most effective tools with or without combination of interventions such as balloon dilation or stent placements. ERCP can also play a transitional role in biliary surgery as a temporary intervention to further strategize the therapeutics of biliary diseases.

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## References

- [1] European Association for the Study of the Liver. EASL Clinical Practice Guidelines: Management of cholestatic liver diseases. *Journal of Hepatology*. 2009;**51**(2):237-267
- [2] McCune WS, Shorb PE, Moscovitz H. Endoscopic cannulation of the ampulla of Vater: A preliminary report. *Annals of Surgery*. 1968;**167**(5):752
- [3] Bali MA, Pezzullo M, Pace E, Morone M. Benign biliary diseases. *European Journal of Radiology*. 2017;**93**:217-228
- [4] NIH Consensus Development Program. NIH state-of-the-science statement on endoscopic retrograde cholangiopancreatography (ERCP) for diagnosis and therapy. National Institutes of Health, Office of the Director. Indiana University; 2002
- [5] Chathadi KV, Chandrasekhara V, Acosta RD, Decker GA, Early DS, Eloubeidi MA, et al. The role of ERCP in benign diseases of the biliary tract. *Gastrointestinal Endoscopy*. 2015;**81**(4):795-803
- [6] Chandrasekhara V, Khashab MA, Muthusamy VR, Acosta RD, Agrawal D, Bruining DH, et al. Adverse events associated with ERCP. *Gastrointestinal Endoscopy*. 2017;**85**(1):32-47
- [7] Costamagna G, Shah SK, Tringali A. Current management of postoperative complications and benign biliary strictures. *Gastrointestinal Endoscopy Clinics*. 2003;**13**(4):635-648
- [8] Gyedu A, Aday-Aboagye K, Badu-Peprah A. Prevalence of cholelithiasis among persons undergoing abdominal ultrasound at the Komfo Anokye Teaching Hospital, Kumasi, Ghana. *African Health Sciences*. 2015;**15**(1):246-252
- [9] European Association for the Study of the Liver. EASL Clinical Practice Guidelines on the prevention, diagnosis and treatment of gallstones. *Journal of Hepatology*. 2016;**65**:146-181
- [10] Buxbaum JL, Fehmi SM, Sultan S, Fishman DS, Qumseya BJ, Cortessis VK, et al. ASGE guideline on the role of endoscopy in the evaluation and management of choledocholithiasis. *Gastrointestinal Endoscopy*. 2019;**89**(6):1075-1105
- [11] Williams EJ, Green J, Beckingham I, Parks R, Martin D, Lombard M. Guidelines on the management of common bile duct stones (CBDS). *Gut*. 1 Jul 2008;**57**(7):1004-1021
- [12] Tse F, Yuan Y. Early Routine Endoscopic Retrograde Cholangiopancreatography (ERCP) Strategy versus Early Conservative Management Strategy in Acute Gallstone Pancreatitis: A Cochrane Systematic Review and Meta-analysis Presidential Poster: 157. *Official journal of the American College of Gastroenterology | ACG*. 1 Oct 2012;**107**:S67
- [13] Maple JT, Ben-Menachem T, Anderson MA, Appalaneni V, Banerjee S, Cash BD, et al. The role of endoscopy in the evaluation of suspected choledocholithiasis. *Gastrointestinal Endoscopy*. 2010;**7**(1):1-9
- [14] Meine GC, Baron TH. Endoscopic papillary large-balloon dilation combined with endoscopic biliary sphincterotomy for the removal of bile duct stones (with video). *Gastrointestinal Endoscopy*. 2011;**74**(5):1119-1126

- [15] Giger UF, Michel JM, Opitz I, Inderbitzin DT, Kocher T, Krähenbühl L, et al. Risk factors for perioperative complications in patients undergoing laparoscopic cholecystectomy: Analysis of 22,953 consecutive cases from the swiss Association of Laparoscopic and Thoracoscopic Surgery database. *Journal of the American College of Surgeons*. 2006;**203**(5):723-728
- [16] Richardson MC, Bell G, Fullarton GM. Incidence and nature of bile duct injuries following laparoscopic cholecystectomy: An audit of 5913 cases. West of Scotland laparoscopic cholecystectomy audit group. *British Journal of Surgery*. 1996;**83**(10):1356-1360
- [17] De Reuver PR, Busch OR, Rauws EA, Lameris JS, Van Gulik TM, Gouma D. Long-term results of a primary end-to-end anastomosis in peroperative detected bile duct injury. *Journal of Gastrointestinal Surgery*. 2007;**11**(3):296-302
- [18] Rauws EA, Gouma DJ. Endoscopic and surgical management of bile duct injury after laparoscopic cholecystectomy. *Best Practice & Research. Clinical Gastroenterology*. 2004;**18**(5):829-846
- [19] Nikpour AM, Knebel RJ, Cheng D. Diagnosis and Management of Postoperative Biliary Leaks. *Seminars in Interventional Radiology*. Dec 2016;**33**(4):307-312. DOI: 10.1055/s-0036-1592324. PMID: 27904250; PMCID: PMC5088090
- [20] Copelan A, Bahoura L, Tardy F, Kirsch M, Sokhandon F, Kapoor B. Etiology, diagnosis, and management of bilomas: a current update. *Techniques in vascular and interventional radiology*. 1 Dec 2015;**18**(4):236-243
- [21] Seeras K, Qasawa RN, Kashyap S, et al. Bile Duct Repair. [Updated 2022 Jun 5]. In: StatPearls [Internet]. Treasure Island, FL: StatPearls Publishing; 2022. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK525989/>
- [22] Shanbhogue AK, Tirumani SH, Prasad SR, Fasih N, McInnes M. Benign biliary strictures: A current comprehensive clinical and imaging review. *American Journal of Roentgenology*. 2011;**197**(2):W295-W306
- [23] Paranandi B, Oppong KW. Biliary strictures: Endoscopic assessment and management. *Frontline Gastroenterology*. 2017;**8**(2):133-137
- [24] Costamagna G, Pandolfi M, Mutignani M, Spada C, Perri V. Long-term results of endoscopic management of postoperative bile duct strictures with increasing numbers of stents. *Gastrointestinal Endoscopy*. 2001;**54**(2):162-168
- [25] Kassab C, Prat F, Liguory C, Meduri B, Ducot B, Fritsch J, et al. Endoscopic management of post-laparoscopic cholecystectomy biliary strictures: Long-term outcome in a multicenter study. *Gastroentérologie Clinique et Biologique*. 2006;**30**(1):124-129
- [26] Zepeda-Gómez S, Baron TH. Benign biliary strictures: Current endoscopic management. *Nature Reviews. Gastroenterology & Hepatology*. 2011;**8**(10):573-581
- [27] Feldman M, Friedman L, Sleisenger M. Sleisenger & Fordtran's Gastro-Intestinal and Liver Disease: Pathophysiology, Diagnosis and Management. 7th ed. Philadelphia: Saunders; 2002. pp. 1131-1137
- [28] Kowdley KV. Primary Sclerosing Cholangitis: Epidemiology and Pathogenesis. Waltham, MA; 2015

- [29] Primary Sclerosing Cholangitis. 2022. Available from: <https://next.amboss.com/us/article/nS07Zf?q=Sclerosing%20cholangitis#Z17ab40a6a2ee805421ace9478cfb909c> [Accessed: August 26, 2022]
- [30] Baluyut AR, Sherman S, Lehman GA, Hoen H, Chalasani N. Impact of endoscopic therapy on the survival of patients with primary sclerosing cholangitis. *Gastrointestinal Endoscopy*. 2001;**53**(3):308-312
- [31] Singh S, Talwalkar JA. Primary sclerosing cholangitis: Diagnosis, prognosis, and management. *Clinical Gastroenterology and Hepatology*. 2013;**11**(8):898-907
- [32] Ghazale A, Chari ST, Zhang L, Smyrk TC, Takahashi N, Levy MJ, et al. Immunoglobulin G4-associated cholangitis: Clinical profile and response to therapy. *Gastroenterology*. 2008;**134**(3):706-715
- [33] Culver EL, Webster G. IgG4-related sclerosing cholangitis. *Biliary Disease*. 2017:243-261
- [34] Skaro AI, Jay CL, Baker TB, Wang E, Pasricha S, Lyuksemburg V, et al. The impact of ischemic cholangiopathy in liver transplantation using donors after cardiac death: The untold story. *Surgery*. 2009;**146**(4):543
- [35] de Buy Wenniger LJ, Culver EL, Beuers U. Exposure to occupational antigens might predispose to IgG4-related disease. *Hepatology (Baltimore, Md.)*. 2014;**60**(4):1453
- [36] Campbell WL, Sheng R, Zajko AB, Abu-Elmagd K, Demetris AJ. Intrahepatic biliary strictures after liver transplantation. *Radiology*. 1994;**191**(3):735-740
- [37] Tung BY, Kimmey MB. Biliary complications of orthotopic liver transplantation. *Digestive Diseases*. 1999;**17**(3):133-144
- [38] Guichelaar MM, Benson JT, Malinchoc M, Krom RA, Wiesner RH, Charlton MR. Risk factors for and clinical course of non-anastomotic biliary strictures after liver transplantation. *American Journal of Transplantation*. 2003;**3**(7):885-890
- [39] Okuno WT, Whitman GJ, Chew FS. Recurrent pyogenic cholangiohepatitis. *American journal of roentgenology*. 1996;**167**(2):484
- [40] Afagh A, Pancu D. Radiologic findings in recurrent pyogenic cholangitis. *The Journal of Emergency Medicine*. 2004;**26**(3):343-346
- [41] Gupta A, Simo K. Recurrent Pyogenic Cholangitis. In: StatPearls [Internet]. Treasure Island, FL: StatPearls Publishing; 9 May 2022. PMID: 33231978
- [42] Jain M, Agarwal A. MRCP findings in recurrent pyogenic cholangitis. *European Journal of Radiology*. 2008;**66**(1):79-83
- [43] Al-Sukhni W, Gallinger S, Pratzler A, Wei A, Ho CS, Kortan P, et al. Recurrent pyogenic cholangitis with hepatolithiasis—The role of surgical therapy in North America. *Journal of Gastrointestinal Surgery*. 2008;**12**(3):496-503
- [44] Yoon HK, Sung KB, Song HY, Kang SG, Kim MH, Lee SG, et al. Benign biliary strictures associated with recurrent pyogenic cholangitis: Treatment with expandable metallic stents. *American Journal of Roentgenology*. 1997;**169**(6):1523-1527
- [45] Waechter FL, Sampaio JA, Pinto RD, Alvares-da-Silva MR, Cardoso FG,



- Francisconi C, et al. The role of liver transplantation in patients with Caroli's disease. *Hepato-Gastroenterology*. 2001;**48**(39):672-674
- [46] Lu SC, Debian KA. Cystic diseases of the biliary tract. In: Yamada T, Alpers DH, Kaplowitz N, Laine L, Owyang C, Powell DW, editors. *Textbook of Gastroenterology*. Philadelphia: Lippincott Williams and Wilkins; 2003. pp. 2225-2233
- [47] Yonem O, Bayraktar Y. Clinical characteristics of Caroli's disease. *World Journal of Gastroenterology*. 2007;**13**(13):1930
- [48] Saxena R, Pradeep R, Chander J, Kumar P, Wig JD, Yadav RV, et al. Benign disease of the common bile duct. *The British Journal of Surgery*. 1988;**75**(8):803-806
- [49] Olbourne NA. Choledochal cysts. A review of the cystic anomalies of the biliary tree. *Annals of the Royal College of Surgeons of England*. 1975;**56**(1):26
- [50] Tan SS, Tan NC, Ibrahim S, Tay KH. Management of adult choledochal cyst. *Singapore Medical Journal*. 2007;**48**(6):524
- [51] Choledochal Cyst. 2022. Available from: <https://radiopaedia.org/articles/choledochal-cyst>. [Accessed August 28, 2022]
- [52] De Angelis P, Foschia F, Romeo E, Caldaro T, Rea F, Di Abriola GF, et al. Role of endoscopic retrograde cholangiopancreatography in diagnosis and management of congenital choledochal cysts: 28 pediatric cases. *Journal of Pediatric Surgery*. 2012;**47**(5):885-888
- [53] Drossman DA, Li Z, Andruzzi E, Temple RD, Talley NJ, Grant Thompson W, et al. US householder survey of functional gastrointestinal disorders. *Digestive Diseases and Sciences*. 1993;**38**(9):1569-1580
- [54] Corazziari E, Shaffer EA, Hogan WJ, Sherman S, Toouli J. Functional disorders of the biliary tract and pancreas. *Gut*. 1999;**45**(2):48-54
- [55] Cheon YK. How to interpret a functional or motility test-sphincter of oddi manometry. *Journal of Neurogastroenterology and Motility*. 2012;**18**(2):211
- [56] Sherman S. What is the role of ERCP in the setting of abdominal pain of pancreatic or biliary origin (suspected sphincter of Oddi dysfunction)? *Gastrointestinal Endoscopy*. 2002;**56**(6):S258-S266
- [57] Kuo WH, Pasricha PJ, Kalloo AN. The role of sphincter of Oddi manometry in the diagnosis and therapy of pancreatic disease. *Gastrointestinal Endoscopy Clinics of North America*. 1998;**8**(1):79-85
- [58] Lim CH, Jahansouz C, Freeman ML, Leslie DB, Ikramuddin S, Amateau SK. Outcomes of endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy for suspected sphincter of Oddi dysfunction (SOD) post Roux-en-Y gastric bypass. *Obesity Surgery*. 2017;**27**(10):2656-2662
- [59] Sherman S, Hawes RH, Madura JA, Lehman GA. Comparison of intraoperative and endoscopic manometry of the sphincter of Oddi. *Surgery, Gynecology & Obstetrics*. 1992;**175**(5):410-418
- [60] Abou-Saif A, Al-Kawas FH. Complications of gallstone disease: Mirizzi syndrome, cholecystocholedochal fistula, and



gallstone ileus. *The American journal of gastroenterology*. 2002;**97**(2):249-254

[61] Curet MJ, Rosendale DE, Congilosi S. Mirizzi syndrome in a native American population. *The American journal of surgery*. 1994;**168**(6):616-621

[62] Tanaka N, Nobori M, Furuya T, Ueno T, Kimura H, Nagai M, et al. Evolution of Mirizzi syndrome with biliobiliary fistula. *Journal of Gastroenterology*. 1995;**30**(1):117-121

[63] Csendes A, Diaz JC, Burdiles P, Maluenda F, Nava O. Mirizzi syndrome and cholecystobiliary fistula: A unifying classification. *Journal of British Surgery*. 1989;**76**(11):1139-1143

[64] Ahlawat SK, Singhanian R, Al-Kawas FH. Mirizzi syndrome. *Current Treatment Options in Gastroenterology*. 2007;**10**(2):102-110

[65] Tan KY, Chng HC, Chen CY, Tan SM, Poh BK, Hoe MN. Mirizzi syndrome: Noteworthy aspects of a retrospective study in one Centre. *ANZ Journal of Surgery*. 2004;**74**(10):833-837

[66] Cruz FO, Barriga P, Tocornal J, Burhenne HJ. Radiology of the Mirizzi syndrome: Diagnostic importance of the transhepatic cholangiogram. *Gastrointestinal Radiology*. 1983;**8**(1):249-253

[67] Ravo B, Epstein H, La Mendola S, Ger R. The Mirizzi syndrome: Preoperative diagnosis by sonography and transhepatic cholangiography. *American Journal of Gastroenterology (Springer Nature)*. 1986;**81**(8)

[68] Khan TF, Muniandy S, Hayat FZ, Sherazi ZA, Nawaz MH. Mirizzi syndrome--a report of 3 cases with a review of the present

classifications. *Singapore Medical Journal*. 1999;**40**(3):171-173

[69] Chowbey PK, Sharma A, Mann V, Khullar R, Baijal M, Vashistha A. The management of Mirizzi syndrome in the laparoscopic era. *Surgical Laparoscopy, Endoscopy & Percutaneous Techniques*. 2000;**10**(1):11-14

[70] Bektaş M, Dökmeci A, Cinar K, Halici I, Oztas E, Karayalcin S, et al. Endoscopic management of biliary parasitic diseases. *Digestive Diseases and Sciences*. 2010;**55**(5):1472-1478

[71] Sandouk F, Haffar S, Zada MM, Graham DY, Anand BS. Pancreaticobiliary ascariasis: Experience of 300 cases. *American Journal of Gastroenterology (Springer Nature)*. 1997;**92**(12)