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Review Article

Surgery Section

Mirizzi Syndrome: A Narrative Review of Classification, Diagnosis and Surgical Management

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ABSTRACT

Mirizzi syndrome is a rare but serious complication of gallstone disease resulting from common hepatic duct obstruction due to either extrinsic compression or inflammatory changes. The Csendes classification is described, which provides a method for assessing the severity of the syndrome and determining whether to undertake surgical intervention. It will also outline possible diagnostic types {(Ultrasonography (USG), Computed Tomography (CT), Magnetic Resonance Cholangiopancreatography (MRCP), Endoscopic Retrograde Cholangiopancreatography (ERCP)} to increase detection rates. Treatment varies by spectrum; in mild presentations, a simple cholecystectomy is sufficient, whereas more significant presentations require biliary reconstruction. This review restates recent advances in minimally invasive and endoscopic techniques that improve patient outcomes in this syndrome. For this reason, healthcare workers need to be aware of Mirizzi syndrome so they can address each case uniquely, prevent various risks during surgery, and ultimately enhance the well-being of patients. This narrative review highlights the medical issues, underlying causes, and factors that increase the risk of the syndrome, noting that diagnosis is challenging because its symptoms are often unclear and similar to those of other biliary disorders. Therefore, the current review is necessary to consolidate current evidence on the pathogenesis, diagnostic challenges, and evolving management strategies of Mirizzi syndrome, thereby guiding clinical decision-making.

Keywords: Biliary obstruction, Cholecystectomy, Diagnostic imaging, Endoscopic techniques, Hepatobiliary disorders

INTRODUCTION

Mirizzi syndrome is a rare but significant complication of chronic cholelithiasis [1,2]. It is caused by either extrinsic compression or erosion of the common hepatic duct by an impacted gallstone in the cystic duct or Hartmann's pouch. [3,4]. Mirizzi syndrome was first described by Pablo Mirizzi in 1948 [1]. The incidence of Mirizzi syndrome among patients undergoing cholecystectomy varies greatly, illustrating an incidence rate ranging from 0.05% to 5.7% [5]. It presents a diagnostic and therapeutic dilemma due to its abnormal presentation of the biliary tree and the proximity of the biliary structures to intervening critical vasculature [1,6]. Patients commonly present with an array of symptoms that overlap with other types of biliary obstruction, including but not limited to: jaundice, right upper quadrant pain, and symptoms of cholangitis [6,7]. If not recognised, morbid complications can occur, including injury to the bile duct during cholecystectomy [8,9].

Mirizzi Syndrome

Mirizzi syndrome is a rare complication of gallstone disease that presents with a variety of symptoms mainly due to the extrinsic compression of the common hepatic duct by impacted gallstones in the cystic duct or neck of the gallbladder [10]. The most common clinical findings are right upper quadrant abdominal pain and jaundice [11]. These symptoms can often be confused with those of other biliary pathologies, making preoperative diagnosis difficult [11,12]. In addition to the classic symptoms, patients may also experience nausea, vomiting, and signs of cholangitis, such as fever and an increased white blood cell count [13]. In rare instances, Mirizzi syndrome may exhibit physiologically without pain, such that the only finding is a painless obstructive jaundice [13,14]. There are cases where the diagnosis is noted in asymptomatic patients whose condition is discovered incidentally through imaging or surgical exploration [14].

Classification and Surgical Approach to Mirizzi Syndrome According to Csendes

Mirizzi syndrome can progress into cholecystobiliary or cholecystoenteric fistulas [15]. The Csendes classification system is the most common way to classify Mirizzi syndrome, and it covers five types based on how much of the bile duct is affected and if a fistula has formed [Table/Fig-1] [16].

Alternative Classification Systems for Mirizzi Syndrome

Beyond the widely used Csendes classification, there are a few other alternative systems of classification for Mirizzi syndrome [17]. The McSherry classification (1982) was the first frequently applied system and divides Mirizzi syndrome into two categories: Type I, denoting external compression of the common hepatic duct by an impacted gallstone, and Type II, indicating the presence of a cholecystobiliary fistula [18]. In 1997, Nagakawa T et al., also introduced a modified classification with four types: Type I and II match McSherry's definitions; Type III describes stones located at the confluence of the cystic duct and common hepatic duct; and Type IV denotes bile duct strictures due to inflammation (e.g., cholecystitis), without stones [19]. Another significant alternative for the classification of Mirizzi syndrome is the Beltrán classification (2012), which further simplified previous systems by restructuring the cholecystobiliary fistula types into IIa (<50% of bile duct diameter) and IIb (>50%), and reclassifying cholecystoenteric fistulas as Type IIIa (without gallstone ileus) and IIIb (with gallstone ileus) [20].

Predisposing Factors and Pathogenesis of Mirizzi Syndrome

Mirizzi syndrome generally develops from chronic cholelithiasis, where chronic inflammation and pressure from the impacted stone lead to erosion of the bile duct and the potential formation of a cholecystobiliary fistula [21]. Central to its pathogenesis is the impaction of a gallstone in the cystic duct or Hartmann's pouch,

Type	Definition	Fistula presence	Extent of bile duct involvement	Surgical management
Type I [16]	External compression of the common hepatic duct by a gallstone impacted in the cystic duct or gallbladder neck	Absent	No erosion of the bile duct	Cholecystectomy ± Choledochostomy
Type II [16]	Cholecystobiliary fistula due to erosion of gallstone into the common hepatic duct	Present	< 1/3 circumference of the common hepatic duct	Fistula repair with absorbable sutures or choledochoplasty using gallbladder remnant
Type III [16]	More extensive cholecystobiliary fistula with erosion into the common hepatic duct	Present	≤2/3 circumference of the common hepatic duct	Choledochoplasty; direct suture repair not advised
Type IV [16]	Complete destruction of the common hepatic duct wall due to fistula	Present	Total circumference of common hepatic duct	Roux-en-Y hepaticojejunostomy (bilioenteric anastomosis)
Type Va [16]	Cholecystoenteric fistula without gallstone ileus	Present	Involves both biliary and enteric systems	Cholecystectomy + Fistula repair
Type Vb [16]	Cholecystoenteric fistula with gallstone ileus	Present	Involves both biliary and enteric systems	Enterolithotomy + Cholecystectomy

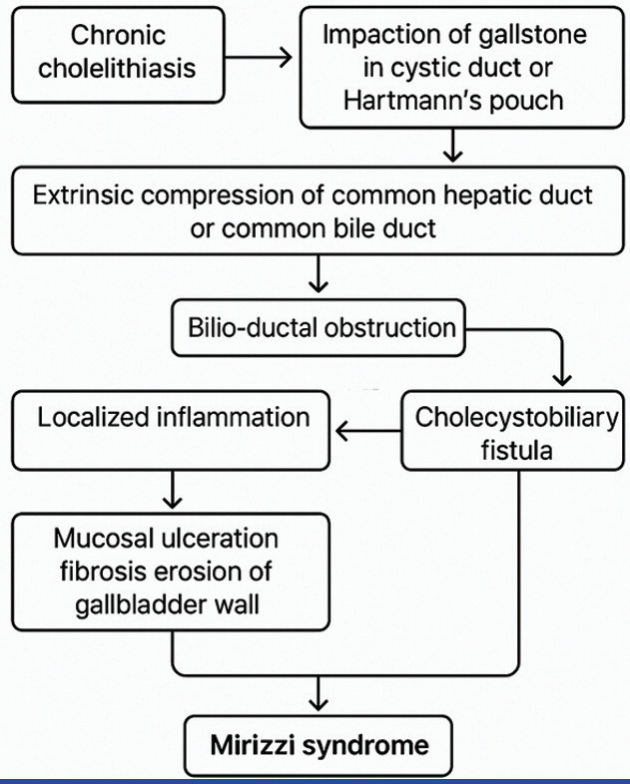
[Table/Fig-1]: Csendes classification of Mirizzi syndrome.

which causes extrinsic compression of the adjacent Common Hepatic Duct (CHD) or Common Bile Duct (CBD) [15,22]. This leads to biliary-ductal obstruction, either partial or complete, manifesting in clinical obstructive jaundice [22]. Over time, the mechanical pressure from the impacted stone leads to a localised inflammatory response, leading to mucosal ulceration, fibrosis, and, in rare cases, erosion of the gallbladder wall into the bile duct [23]. The above can develop into a cholecystocholedochal or cholecystohepatic fistula, which is more notable in more advanced forms of the syndrome [23,24]. Long-standing gallstone disease is the leading risk factor for developing Mirizzi syndrome [24]. Large gallstones and multiple gallstones increase the risk of cystic duct obstruction or Hartmann's pouch obstruction due to biliary anatomy when the biliary anatomy has narrow or apposed anatomy [24]. When anatomical variation occurs, such as a low insertion of the cystic duct or a parallel cystic duct to the common hepatic duct, it creates a predisposition due to increased anatomical contact and mechanical stress on the structures [25].

Delayed operative management with symptomatic cholelithiasis is an additional risk factor, as is recurrent cholecystitis and older age [25,26]. These risks enhance the duration and severity of inflammation, thus allowing fibrosis and the anatomical distortion of the biliary structures [26]. It is also noted that Mirizzi syndrome occurs more frequently in women, likely because gallstone disease occurs more often in females [27]. In addition, patients presenting from geographic regions with limited access to early surgical care and imaging present with advanced Mirizzi syndrome, thus increasing the complication rate of fistula [26]. The pathogenesis of Mirizzi syndrome is depicted in [Table/Fig-2].

Diagnostic Modalities in Mirizzi Syndrome

Due to its non specific clinical presentation and often misleading appearance that mimics other forms of hepatobiliary illness, diagnosing Mirizzi syndrome, a rare complication of gallstone disease, requires a high degree of suspicion [28]. Patients typically present with right upper quadrant pain, fever, and jaundice, which may resemble acute cholangitis or obstructive jaundice



[Table/Fig-2]: Pathogenesis of Mirizzi syndrome.

[29]. Laboratory analysis usually shows a cholestatic liver enzyme pattern with elevated bilirubin, alkaline phosphatase, and gamma-glutamyl transpeptidase levels [29,30]. However, these results are not pathognomonic and must be interpreted in conjunction with imaging studies [30].

Imaging is a key component in making the diagnosis. Ultrasound of the abdomen is the most frequently used modality and will show gallstones and intrahepatic duct enlargement, but has poor sensitivity regarding extrinsic compression of the common hepatic duct [31]. Computed Tomography (CT) will give better anatomical detail and can also be used to suggest the presence of malignancy [32]. Magnetic Resonance Cholangiopancreatography (MRCP) is the non invasive gold standard imaging modality of choice; it visualises the biliary anatomy at the obstructed level and causes [33]. Endoscopic Retrograde Cholangiopancreatography (ERCP) is both a diagnostic and therapeutic modality and visualises the biliary tree, identifies the fistula, and stents in an attempt to relieve obstruction [28,34]. In some severe cases, laparoscopy can also help diagnose the problem, especially if regular imaging does not give a definite result or there is a suspicion of cancer [15].

Differentiating Mirizzi Syndrome from Mimicking Hepatobiliary Pathologies

Since Mirizzi syndrome has symptoms much like choledocholithiasis, cholangiocarcinoma, gallbladder carcinoma and bile duct strictures, it must be correctly identified for proper treatment [35]. Mirizzi syndrome is caused by external pressure on the duct, while choledocholithiasis usually results from stones lodged within it [36]. Often, intraductal stones with resulting proximal dilation are seen on MRCP or Endoscopic Ultrasound (EUS) [36]. Choledocholithiasis is often diagnosed by visualising filling defects within the duct lumen, and ERCP [37].

Cholangiocarcinoma is a malignant neoplasm arising from bile duct epithelium, can also present with jaundice and bile duct obstruction [38]. It is characterised by irregular ductal wall thickening and mass lesions, with delayed contrast enhancement on imaging {CT or Magnetic Resonance Imaging (MRI) [39]. This malignancy can often present as asymmetric biliary strictures and elevation in tumour markers (i.e., CA 19-9) [40]. Gallbladder carcinoma commonly

presents as a mass that usually replaces or infiltrates the gallbladder wall, with or without direct invasion of the biliary tree, and on imaging resembles an irregular mass with disruption of the normal gallbladder anatomy, and potentially regional lymphadenopathy [41]. It requires imaging studies such as CT and sometimes Positron Emission Tomography (PET) or Contrast-enhanced CT (CECT) because it may often delay diagnosis; these imaging studies are valuable in aiding differentiation of the Mirizzi syndrome accordingly [41].

Benign bile duct strictures typically occur after surgery, a traumatic event, or in the setting of chronic inflammation; malignant strictures can occur from tumours such as cholangiocarcinoma or pancreatic cancer [42,43]. ERCP with brush cytology and cholangioscopy assists in differentiation; strictures tend to cause longer segment narrowing and lack the associated gallstones, causing external compression [42]. Comparative differentiation of Mirizzi syndrome from other hepatobiliary pathologies is mentioned in [Table/Fig-3].

Contemporary and Emerging Approaches to the Management of Mirizzi Syndrome

In the early stages of the Mirizzi syndrome, where there is external compression of the common hepatic duct but no cholecystocholedochal fistula, laparoscopic or open cholecystectomy with or without choleductal cleaning may be sufficient [44]. However, in practice, during laparoscopic cholecystectomy, the surgeon must navigate through significant inflammation and/or adhesive disease, which may prevent them from completing the procedure laparoscopically and may increase the risk for bile duct injury. In such cases, converting the laparoscopic approach to an open approach to allow for safe dissection and complete removal of the gallbladder is suggested [45]. In advanced Mirizzi syndrome, where cholecystocholedochal fistulas are present, more complex biliary reconstruction is needed [46]. Surgical options for repair of these types of defects include partial cholecystectomy with repair of the bile duct defect by T-tube drainage as well as bilioenteric anastomosis (for example, hepaticojejunostomy in more severe cases) [47,48]. Preoperative ERCP with stent placement may be a surgical option for biliary decompression, especially in patients who have cholangitis or are not fit for surgery, although in such cases, ERCP is not an adequate treatment [49].

The treatment strategies for Mirizzi syndrome are progressively using minimally invasive strategies, and further utilising cutting-edge endoscopic techniques to enhance positive outcomes and eliminate surgical risk [50,51]. One important advancement noted is Single Operator Peroral Cholangioscopy (SOPC) with Electrohydraulic Lithotripsy (EHL) [51,52]. As technically feasible, SOPC allows visualisation and fragmentation of impacted stones. SOPC has shown >90% success rates for patients with complicated conditions, including cholecystocholedochal fistula, and offers less surgical intervention [52].

Building on these advances, current management shows clear strengths but also important limitations and real-world challenges [53]. Strengths include expanding minimally invasive armamentarium; laparoscopic subtotal "fundus-first" or intra-cholecystic approaches that can safely control inflammation while minimising ductal injury when anatomy is hostile, and "one-session" strategies that combine laparoscopic cholecystectomy with bile-duct exploration to clear stones and shorten care pathways in selected patients [53]. Likewise, digital SOPC, along with electrohydraulic (or laser) lithotripsy, offers direct visualisation and high stone-clearance rates even in complex Mirizzi subtypes, reducing the need for extensive open reconstruction.

Management approaches for Mirizzi syndrome are described in [Table/Fig-4].

Current Gaps and Future Directions in Mirizzi Syndrome

The existing literature on Mirizzi syndrome is limited by heterogeneous diagnostic pathways, fragmented and inconsistently validated classification systems, and predominantly small, retrospective, single-centre studies, which focus particularly on emerging minimally invasive approaches, such as single-operator cholangioscopy with lithotripsy [52,54]. Preoperative diagnosis remains variable despite advances in MRCP/ERCP, and no consensus exists on an optimal imaging algorithm or on which classification best predicts surgical complexity and outcomes [29]. Management evidence lacks robust comparative and long-term data, especially from low-resource settings [20].

Features	Mirizzi syndrome [35]	Choledocholithiasis [36,37]	Cholangiocarcinoma [38,39]	Gallbladder carcinoma [41]	Bile duct strictures [42,43]
Etiology	Extrinsic compression of the bile duct by an impacted gallstone in the cystic duct/gallbladder	Stone(s) lodged within the CBD	Malignant tumour of the bile duct epithelium	Malignant tumour arising from gallbladder epithelium	Inflammation, fibrosis (benign), or malignancy causing bile duct narrowing
Clinical presentation	Jaundice, RUQ pain, sometimes cholangitis	Jaundice, biliary colic, cholangitis	Painless jaundice, weight loss, pruritus	Jaundice, weight loss, RUQ mass	Jaundice, cholangitis, history of biliary surgery or pancreatitis
Imaging findings	Extrinsic compression of bile duct; impacted stone at neck/cystic duct	Intraluminal filling defects in CBD on MRCP/ERCP	Irregular ductal wall thickening, asymmetric stricture, delayed enhancement on CT/MRI	Mass replacing/infiltrating gallbladder, disrupted anatomy, regional lymphadenopathy	Focal luminal narrowing with proximal dilation; longer segment narrowing
Diagnostic tools	Ultrasound, MRCP, ERCP, EUS	MRCP, ERCP (diagnostic and therapeutic)	CT, MRI, ERCP with brush cytology, biopsy	CT, MRI, PET; histopathology confirms diagnosis	ERCP with brush cytology, cholangioscopy, biopsy
Histopathology	Not needed for diagnosis; imaging is often sufficient	Not typically required	Required to confirm diagnosis	Required to confirm diagnosis	Required to rule out malignancy
Tumour markers	Normal	Normal	↑ CA (Carbohydrate Antigen) 19-9, Carcinoembryonic Antigen (CEA)	↑ CA 19-9, CEA	May be elevated if malignant
Therapeutic approach	Often requires surgical intervention (e.g., cholecystectomy)	Endoscopic stone removal via ERCP	Oncologic resection, chemotherapy, palliative care	Surgical resection if early; palliative care in advanced cases	Depends on cause: dilatation (benign), surgery, stenting, or oncologic therapy
Presence of gallstones	Yes	Yes	No	Sometimes	No
Compression vs. infiltration	Extrinsic compression	Intraluminal obstruction	Infiltrative mass lesion	Infiltrative or mass-forming lesion	Narrowing due to fibrosis or inflammation

[Table/Fig-3]: Comparative differentiation of Mirizzi syndrome from other hepatobiliary pathologies[35-39,41-43].

Type (Csendes)	Surgical approach	Procedure	Outcome	Complications
Type I (External compression of common hepatic duct, no fistula)	Laparoscopic cholecystectomy (preferred) or open cholecystectomy if severe inflammation/adhesions	Cholecystectomy ± choledochal cleaning	Good outcomes if anatomy clearly defined; avoids fistula progression	Risk of bile duct injury; conversion to open surgery if inflammation/adhesions severe
Type II–IV (Cholecystocholedochal fistula)	Open or advanced laparoscopic surgery by experienced hepatobiliary surgeon	Partial cholecystectomy + bile duct repair over T-tube (mild defects) OR bilioenteric anastomosis (e.g., hepaticojejunostomy) in severe cases	Restores biliary continuity; prevents recurrent cholangitis	Risk of bile duct stricture; re-intervention possible in T-tube repairs
Any type with cholangitis or high surgical risk	ERCP ± preoperative stenting	Biliary decompression, stone retrieval (if possible)	Symptom relief; bridge to surgery	Not definitive treatment; recurrence if definitive surgery not done
Advanced endoscopic option (all types if technically feasible)	SOPC+EHL	Direct visualisation + stone fragmentation; may combine with laparoscopic cholecystectomy	>90% success in complex cases; less invasive than open reconstruction	Requires advanced expertise and equipment; not universally available
Minimally invasive advanced strategy	Laparoscopic subtotal "fundus-first" or intra-cholecystic approach ± one-session bile duct exploration	Safe dissection in hostile anatomy; stone clearance in same setting	Minimises ductal injury; shortens hospital stay	Requires skilled team; still risk of conversion and bile duct injury

[Table/Fig-4]: Management approaches for Mirizzi syndrome.

CONCLUSION(S)

Mirizzi syndrome is a rare but essential complication of gallstone disease. Correct preoperative identification of Mirizzi syndrome is difficult due to the variable clinical presentation and overlap with other hepatobiliary diseases. The critical issue in management is early identification of Mirizzi syndrome to prevent complications such as injury to the bile duct. Improved imaging modalities and advancements in minimally invasive and endoscopic techniques have reduced diagnostic delays and improved subsequent outcomes.

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