Mirizzi syndrome

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INTRODUCTION

Mirizzi syndrome is defined as common hepatic duct obstruction caused by extrinsic compression from an impacted stone in the cystic duct or infundibulum of the gallbladder [1-3]. Patients with Mirizzi syndrome can present with jaundice, fever, and right upper quadrant pain. Mirizzi syndrome is often not recognized preoperatively in patients undergoing cholecystectomy and can lead to significant morbidity and biliary injury, particularly with laparoscopic surgery [4].

This topic reviews the epidemiology, clinical manifestations, diagnosis, and management of Mirizzi syndrome. Other complications of gallstone disease including choledocholithiasis, acute cholangitis, and acute cholecystitis are discussed separately. (See "Choledocholithiasis: Clinical manifestations, diagnosis, and management" and "Acute cholangitis: Clinical manifestations, diagnosis, and management" and "Acute calculous cholecystitis: Clinical features and diagnosis".)

EPIDEMIOLOGY

Prevalence — Mirizzi syndrome is estimated to occur in 0.05 to 4 percent of patients undergoing surgery for cholelithiasis [5-8]. Approximately 50 to 77 percent of patients with Mirizzi syndrome are women, which may in part be due to a higher incidence of gallstones in

women. (See "Gallstones: Epidemiology, risk factors and prevention", section on 'Epidemiology'.)

Association with gallbladder cancer — Mirizzi syndrome has been associated with gallbladder cancer [7,9]. It has been hypothesized that recurrent inflammation and biliary stasis may predispose to both conditions. The reported prevalence of gallbladder cancer in patients with Mirizzi syndrome undergoing cholecystectomy ranges from 5 to 28 percent [7,9]. In a retrospective study of 4800 patients who underwent cholecystectomy, Mirizzi syndrome was present in 133 patients, of whom seven (5 percent) had gallbladder cancer [9]. A preoperative diagnosis of gallbladder cancer was made in only one of seven patients. Gallbladder cancer was detected intraoperatively in one patient and only on pathologic examination of the gallbladder in five patients.

PATHOPHYSIOLOGY

The gallbladder consists of the fundus, body, infundibulum, and neck. The body extends from the fundus into the tapered portion, or neck. The neck usually forms a gentle curve, the convexity of which forms the infundibulum, or Hartmann's pouch. The gallbladder is connected at its neck to the cystic duct which empties into the common bile duct. Large gallstones can become impacted in the cystic duct or the infundibulum (figure 1). These stones can produce common hepatic duct obstruction by mechanical obstruction of the hepatic duct because of the proximity of the cystic duct and the common hepatic duct, and secondary inflammation with frequent episodes of cholangitis [10-12]. In rare cases, chronic inflammation may result in bile duct wall necrosis and erosion of the anterior or lateral wall of the common bile duct by impacted stones leading to cholecystobiliary (cholecystohepatic or cholecystocholedochal) fistula.

CLASSIFICATION

Mirizzi syndrome has been classified based on the presence and extent of a cholecystobiliary fistula [13]:

- Type I (11 percent of Mirizzi syndrome): External compression of the common hepatic duct due to a stone impacted at the neck/infundibulum of the gallbladder or at the cystic duct (figure 2).
- Type II (41 percent of Mirizzi syndrome): The fistula involves less than one-third of the circumference of the common bile duct.

- Type III (44 percent of Mirizzi syndrome): Involvement of between one-third and two-thirds of the circumference of the common bile duct.

- Type IV (4 percent of Mirizzi syndrome): Destruction of the entire wall of the common bile duct.

CLINICAL FEATURES

Clinical manifestations — Patients with Mirizzi syndrome can present with jaundice, fever, and right upper quadrant pain. However, all three symptoms are only present in 44 to 71 percent of patients [14]. Pain is the most common presenting feature (54 to 100 percent), followed by jaundice (24 to 100 percent) and cholangitis (6 to 35 percent). Up to one-third of patients have acute cholecystitis on presentation, and in rare cases, acute pancreatitis [15,16]. (See "Clinical manifestations and diagnosis of acute pancreatitis", section on 'Clinical features' and "Acute calculous cholecystitis: Clinical features and diagnosis", section on 'Clinical manifestations'.)

Laboratory findings — The major laboratory findings are elevations in the serum concentrations of alkaline phosphatase and bilirubin in over 90 percent of patients [17,18]. Patients with concurrent acute cholecystitis, cholangitis, or pancreatitis may have leukocytosis.

DIAGNOSIS

Mirizzi syndrome should be suspected in any patients with right upper quadrant pain, jaundice, and fever. The diagnosis of Mirizzi syndrome requires the presence of the following on abdominal imaging (eg, transabdominal ultrasonography, contrast-enhanced computed tomography [CT], magnetic resonance cholangiopancreatogram [MRCP]):

- Dilatation of the biliary system above the level of the gallbladder neck
- The presence of a stone impacted in the gallbladder neck
- An abrupt change to a normal diameter of the common duct below the level of the stone

Diagnostic imaging — Initial diagnostic evaluation in a patient suspected to have Mirizzi syndrome usually begins with abdominal ultrasound or CT. In patients with suspected Mirizzi syndrome based on initial imaging or signs and symptoms suggestive of biliary obstruction (right upper quadrant or epigastric pain, elevated liver biochemical tests), we perform magnetic resonance cholangiopancreatogram to establish the diagnosis of Mirizzi syndrome.

- **Abdominal ultrasound** — Ultrasonography in patients with Mirizzi syndrome generally reveals gallstones and a contracted gallbladder. Features suggestive of Mirizzi syndrome include (image 1) [19]:

  - Dilatation of the biliary system above the level of the gallbladder neck
  - The presence of a stone impacted in the gallbladder neck
  - An abrupt change to a normal width of the common duct below the level of the stone

  However, the diagnosis of Mirizzi syndrome is often difficult because of the close proximity of the cystic and the common bile duct. The sensitivity of abdominal ultrasound in the diagnosis of Mirizzi syndrome is 23 to 46 percent [19].

- **Abdominal CT scan** — Abdominal CT scan can ascertain if malignancy is present by revealing enlarged porta hepatis lymph nodes or hepatic infiltration or metastases [20]. However, it does not significantly add to the diagnostic yield of abdominal ultrasound in identifying the cause of biliary obstruction. In one study, the sensitivity and specificity of abdominal CT for the diagnosis of Mirizzi syndrome were 42 and 99 percent, respectively [21].

- **Magnetic resonance cholangiopancreatogram (MRCP)** — MRCP has a high sensitivity for Mirizzi syndrome. It can also determine the extent of pericholecystic inflammation and aid in the differentiation of Mirizzi syndrome from other gallbladder pathologies such as gallbladder malignancy (image 2 and image 3) [21]. T2 weighted MRCP images can differentiate between a neoplastic and inflammatory mass which may not be possible on abdominal ultrasound or CT scan [22].

Evaluation for a cholecystobiliary fistula — We perform endoscopic retrograde cholangiopancreatography (ERCP) to confirm the diagnosis of Mirizzi syndrome and determine if a cholecystobiliary fistula is present [19]. Percutaneous transhepatic cholangiography (PTC) is typically reserved for patients who are not candidates for ERCP, who have failed ERCP, or who have surgically altered anatomy preventing endoscopic access to the biliary tree.

ERCP and sphincterotomy allows for biliary decompression by internal stenting in patients with obstructive jaundice or cholangitis [5]. ERCP can identify a low-lying cystic duct that might be missed by PTC. Unlike PTC, ERCP can visualize the distal common bile duct in the presence of significant obstruction of the common hepatic duct.

Diagnostic findings of (image 4) Mirizzi syndrome on cholangiography are an eccentric or excavating defect on lateral wall of the common bile duct at the level of cystic duct or gallbladder neck. A stone in the neck of the gallbladder or cystic duct and the gallbladder may

be visualized. Cholecystobiliary fistula can be demonstrated on cholangiography by the passage of contrast material from the proximal dilated biliary channels into the gallbladder [5].

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of Mirizzi syndrome depends on the clinical presentation.

- **Choledocholithiasis** — Patients with choledocholithiasis typically present with fever, jaundice, and right upper quadrant pain, but subacutely with prolonged episodes of pain. Choledocholithiasis can typically be differentiated from Mirizzi syndrome based on abdominal imaging. (See "Choledocholithiasis: Clinical manifestations, diagnosis, and management", section on 'Transabdominal ultrasound'.)

- **Acute cholecystitis** — Patients with acute cholecystitis may have right upper quadrant pain that starts more suddenly. However, patients with acute cholecystitis should not have a significantly elevated bilirubin or alkaline phosphatase unless there is a secondary process causing cholestasis. In addition, abdominal imaging in acute cholecystitis typically reveals a normal common bile duct, gallbladder wall thickening, and a sonographic Murphy's sign. (See "Acute calculous cholecystitis: Clinical features and diagnosis", section on 'Diagnostic approach'.)

There are numerous causes of jaundice in addition to Mirizzi syndrome (table 1). Mirizzi syndrome is differentiated from these other conditions by the presence of biliary-type pain and by a dilated common bile duct on abdominal imaging.

The differential diagnosis of fever and abdominal pain includes:

- Appendicitis (see "Acute appendicitis in adults: Clinical manifestations and differential diagnosis")
- Pancreatitis (see "Clinical manifestations and diagnosis of acute pancreatitis", section on 'Diagnosis')
- Liver abscess (see "Pyogenic liver abscess", section on 'Diagnosis')
- Infected choledochal cysts (see "Biliary cysts", section on 'Clinical manifestations')
- Acute cholangitis (see "Acute cholangitis: Clinical manifestations, diagnosis, and management", section on 'Epidemiology and risk factors')
- Intestinal perforation
- Right lower lobe pneumonia/empyema (see "Clinical evaluation and diagnostic testing for community-acquired pneumonia in adults")

Typically, these entities can be differentiated from Mirizzi syndrome based on the clinical history, laboratory tests, and findings on abdominal imaging (eg, magnetic resonance cholangiopancreatography).

MANAGEMENT

General approach — Surgery is the mainstay of therapy for Mirizzi syndrome, permitting removal of the causal factors: the inflamed gallbladder and the impacted stone. (See 'Surgery' below.)

If the diagnosis of Mirizzi syndrome is made preoperatively, endoscopic retrograde cholangiopancreatography can be both diagnostic and therapeutic as a temporizing measure before surgery, as stenting across the obstruction allows decompression of the common bile duct in patients with obstructive jaundice or cholangitis. Endoscopic removal of common bile duct stones may eliminate the need for common bile duct exploration at the time of surgery.

If Mirizzi syndrome is diagnosed incidentally at the time of cholecystectomy, intraoperative cholangiogram should be performed prior to cholecystectomy to confirm the diagnosis and characterize the biliary anatomy.

For patients who are unsuitable surgical candidates, endoscopic retrograde cholangiopancreatography with stenting can be definitive treatment for Mirizzi syndrome [18,23,24]. (See 'Endoscopic therapy' below.)

Surgery — The surgical approach to Mirizzi syndrome is based on the presence and type of cholecystobiliary fistula [13,25-27]. (See 'Evaluation for a cholecystobiliary fistula' above.)

- Type I: Partial or total cholecystectomy, either laparoscopic or open. Common bile duct exploration is typically not required.
- Type II: Cholecystectomy plus closure of the fistula, either by suture repair with absorbable material, T tube placement, or choledochoplasty with the remnant gallbladder.
- Type III: Choledochoplasty or bilioenteric anastomosis (choledochoduodenostomy, cholecystoduodenostomy, or choledochojejunostomy) depending on the size of the fistula. Suture of the fistula is not indicated.
- Type IV: Bilioenteric anastomosis, typically choledochojejunostomy, is preferred because the entire wall of the common bile duct has been destroyed.

In patients with a cholecystobiliary fistula, exploration of the common duct should be carried out to rule out concomitant choledocholithiasis, unless this has been performed endoscopically [14]. The surgeon should maintain a high index of suspicion for gallbladder cancer. A frozen section should be performed of any suspicious areas of the gallbladder. The management of gallbladder cancer is discussed in detail, separately. (See "Surgical management of gallbladder cancer".)

In patients with type I anatomy, dissection can initially be performed laparoscopically with the plan to convert to an open procedure if necessary [28]. In patients with type II, III, and IV anatomy, a laparoscopic approach is possible, but an open procedure may be required. Laparoscopic management of Mirizzi syndrome can present a challenge because the dense adhesions and edematous inflammatory tissue can cause distortion of the normal anatomy and increase the risk for biliary injury [4]. Laparoscopic surgery as the primary treatment of Mirizzi syndrome is controversial but has been reported in patients with type I and II Mirizzi anatomy [26,28-32]. A systematic review of 10 case series reported that laparoscopic cholecystectomy was successful for 73 of 124 patients (59 percent) with Mirizzi syndrome [33]. In a retrospective series of 35 patients who had surgery for Mirizzi syndrome, laparoscopic surgery was attempted in 15 patients and the conversion rate to open surgery was 67 percent [31]. (See "Complications of laparoscopic cholecystectomy".)

Endoscopic therapy — Endoscopic retrograde cholangiopancreatography and sphincterotomy allows for biliary decompression by internal stenting in patients with obstructive jaundice or cholangitis. Other endoscopic approaches (eg, lithotripsy) can be technically challenging, are less commonly successful, and have been associated with complications. There are also limited data on the effectiveness of this approach [18]. In one series of 25 patients with cholangiographic evidence of Mirizzi syndrome, 12 were referred for surgery after preliminary endoscopic therapy and 13 were treated solely with endoscopy [24]. The endoscopic approach included sphincterotomy, mechanical lithotripsy, dissolution therapy with methyl tert-butyl ether, extracorporeal shockwave lithotripsy, and common bile duct stents. Complete clearance of stones from the biliary tree was achieved in only 3 of 13 patients while nine patients were treated with long-term stenting. Five patients died, including two who died of biliary causes. Long-term success appears to be most likely in patients with type II disease who do not have residual gallbladder stones [34].

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Biliary infection and

obstruction".)

SUMMARY AND RECOMMENDATIONS

- Mirizzi syndrome is defined as common hepatic duct obstruction caused by an extrinsic compression from an impacted stone in the cystic duct or gallbladder neck/infundibulum. (See 'Introduction' above.)

- Mirizzi syndrome is diagnosed in 0.05 to 4 percent of patients undergoing surgery for cholelithiasis. (See 'Epidemiology' above.)

- Mirizzi syndrome results from mechanical obstruction of the hepatic duct because of the proximity of the cystic duct and the common hepatic duct, and secondary inflammation which results in biliary obstruction and possible cholangitis. (See 'Pathophysiology' above.)

- Patients can present with jaundice, fever, and right upper quadrant pain. However, all three symptoms are only present in 44 to 71 percent of patients. Pain is the most common presenting feature. The major laboratory findings are elevations in the serum concentrations of alkaline phosphatase and bilirubin. (See 'Clinical manifestations' above.)

- Mirizzi syndrome should be suspected in patients with right upper quadrant pain, jaundice, and fever. The diagnosis of Mirizzi syndrome requires the presence of the following on abdominal imaging (eg, transabdominal ultrasonography, contrast-enhanced computed tomography [CT], magnetic resonance cholangiopancreatogram [MRCP]) (see 'Diagnosis' above):

  - Dilatation of the biliary system above the level of the gallbladder neck
  - The presence of a stone impacted in the gallbladder neck
  - An abrupt change to a normal diameter of the common duct below the level of the stone

- Initial diagnostic evaluation in a patient with suspected Mirizzi syndrome usually begins with abdominal ultrasound or CT. In patients with suspected Mirizzi syndrome based on initial imaging or signs and symptoms suggestive of biliary obstruction (right upper quadrant or epigastric pain, elevated liver biochemical tests), we perform MRCP to establish the diagnosis of Mirizzi syndrome. We perform endoscopic retrograde cholangiopancreatography (ERCP) to confirm the diagnosis of Mirizzi syndrome and determine if a cholecystobiliary fistula is present. (See 'Diagnosis' above and 'Classification' above.)

- Surgery is the mainstay of therapy for Mirizzi syndrome, permitting removal of the causal factors: the inflamed gallbladder and the impacted stone. The surgical approach to Mirizzi syndrome is based on the presence and type of cholecystobiliary fistula.

  If the diagnosis of Mirizzi syndrome is made preoperatively, ERCP can be both diagnostic and therapeutic as a temporizing measure before surgery in patients with obstructive jaundice or cholangitis. If Mirizzi syndrome is diagnosed incidentally at the time of cholecystectomy, intraoperative cholangiogram should be performed prior to cholecystectomy to confirm the diagnosis and characterize the biliary anatomy. (See 'General approach' above and 'Surgery' above.)

- Endoscopic treatment can be effective as a temporizing measure before surgery and can be definitive treatment for poor surgical candidates. (See 'Endoscopic therapy' above.)

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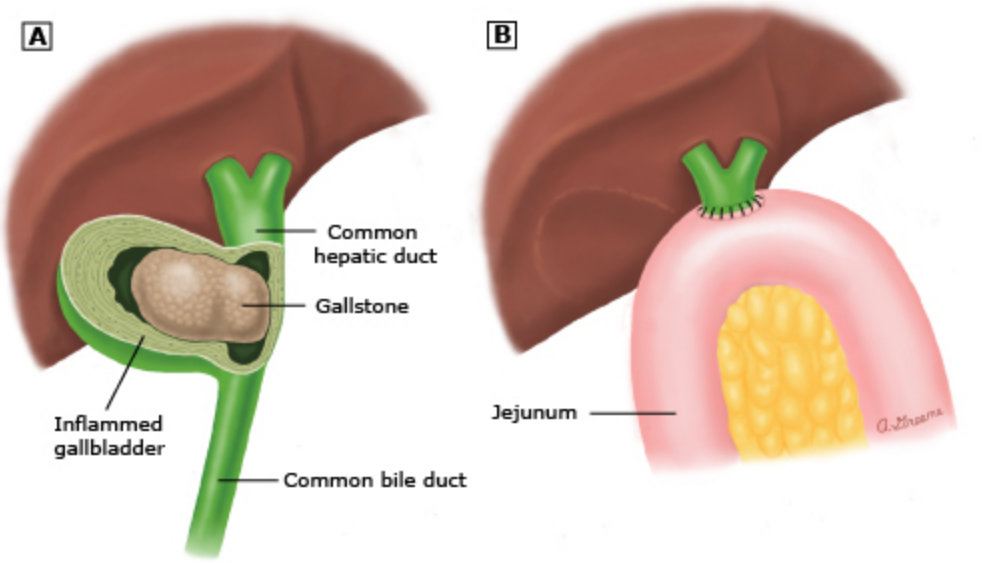
GRAPHICS

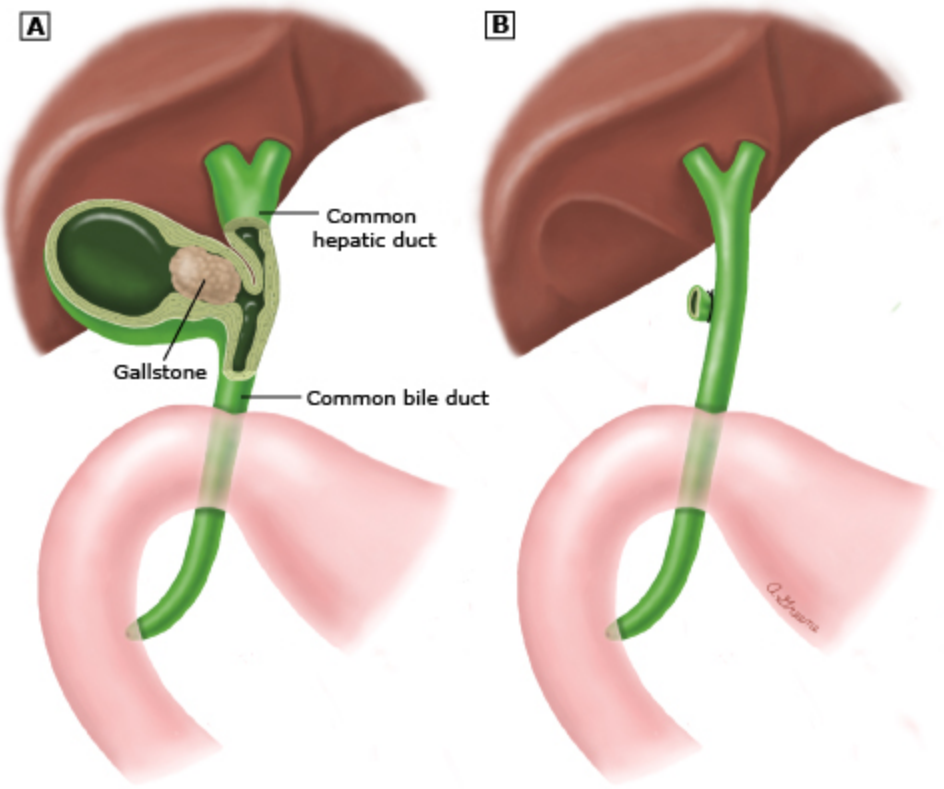
Mirizzi syndrome with erosion of the stone into the common hepatic duct



Surgery is the mainstay of therapy for Mirizzi syndrome, permitting removal of the causal factors: the inflamed gallbladder and the impacted stone. When the stone has eroded into the common bile duct or common hepatic duct, a bilioenteric anastomosis is typically required.

Graphic 54508 Version 3.0

Mirizzi syndrome with compression of the common hepatic duct by an impacted stone



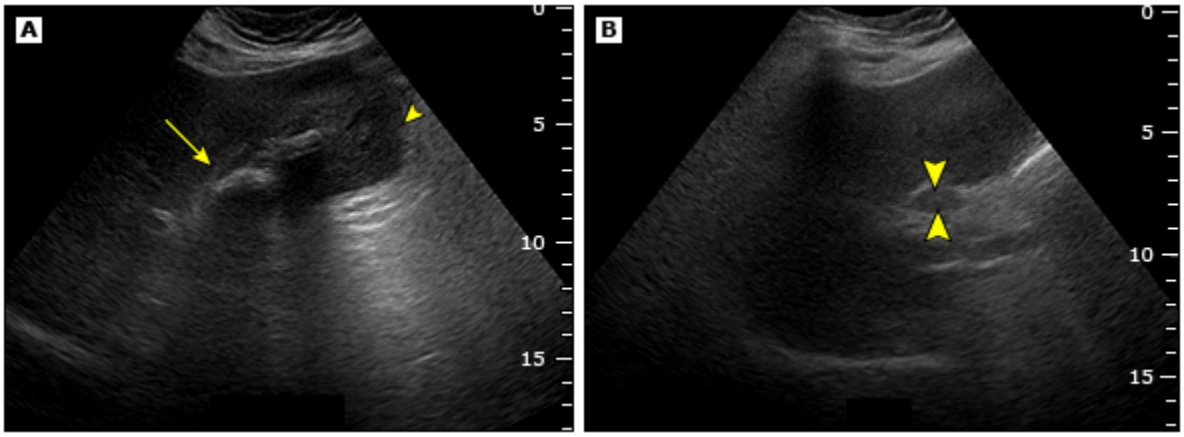
Schematic representation of Mirizzi syndrome.

(Panel A) An impacted stone in the cystic duct or Hartmann's pouch of the gallbladder obstructs the common hepatic duct both by extrinsic compression and associated inflammation.

(Panel B) Treatment consists of cholecystectomy.

Graphic 54888 Version 6.0

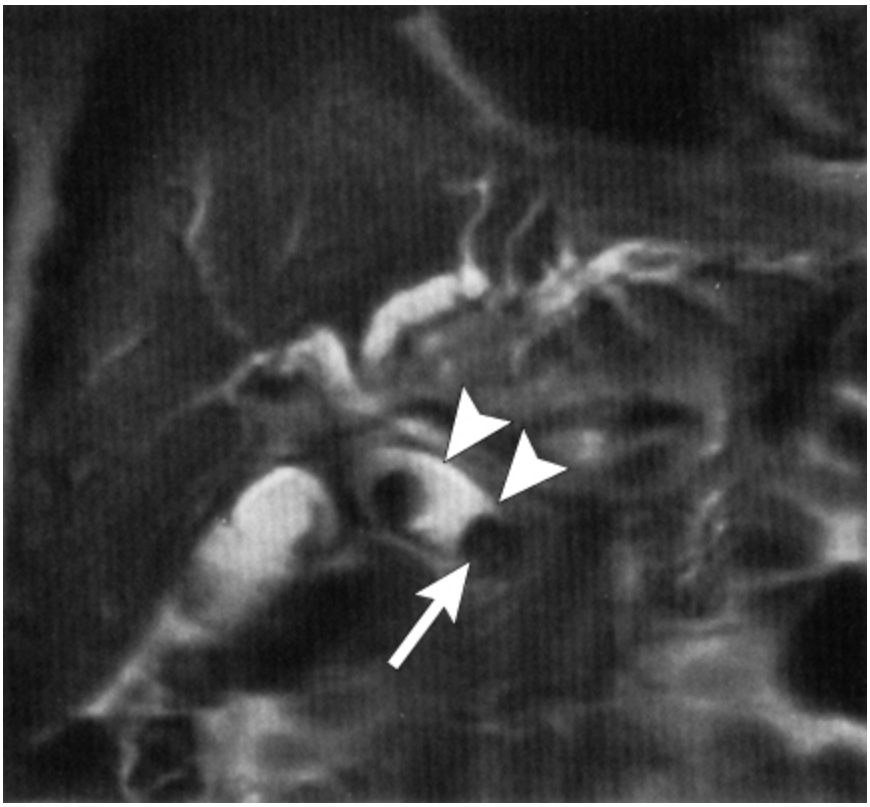
Ultrasound from a patient with acute cholecystitis and Mirizzi syndrome



Transabdominal ultrasound from a patient with acute cholecystitis and Mirizzi syndrome. The patient presented with right upper quadrant pain and jaundice. Acute cholecystitis and Mirizzi syndrome are confirmed by the ultrasound findings of a positive sonographic Murphy's sign (pain with compression of the gallbladder by the ultrasound probe), a large shadowing stone impacted in the infundibulum of the gallbladder (arrow), cholestasis with sludge (small arrowhead), and a dilated common hepatic duct (large arrowheads in B).

Graphic 86878 Version 2.0

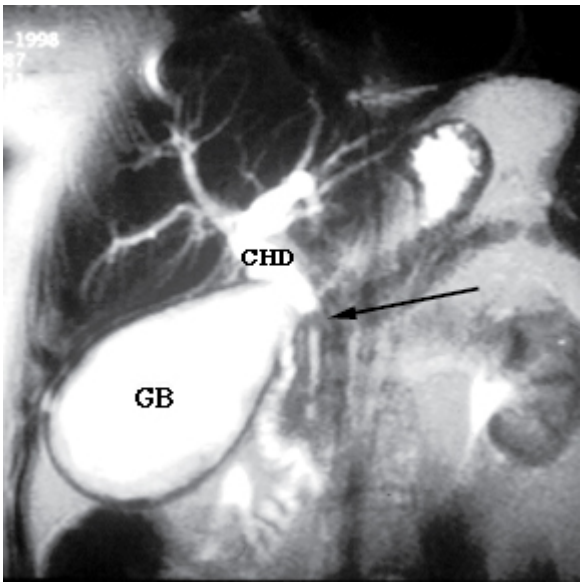
Mirizzi syndrome seen on magnetic resonance cholangiopancreatography (MRCP)



Mirizzi syndrome. Two calculi in the dilated cystic duct (arrowheads), which parallels the extrahepatic bile duct. The inferior calculus (arrow) eroded through the wall of the cystic duct into the extrahepatic bile duct, bridging the two structures and resulting in obstruction of the bile duct.

Graphic 75454 Version 4.0

Mirizzi syndrome seen on magentic resonance cholangiopancreatography (MRCP)

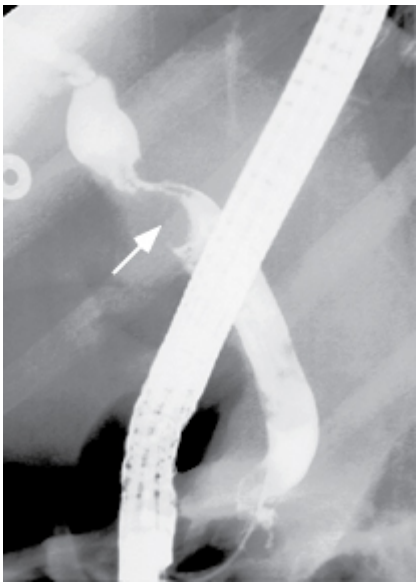


MRCP study in an 81-year-old woman with jaundice and right upper quadrant pain shows dilation of the common hepatic duct (CHD) and gallbladder (GB) with a filling defect in the common bile duct (arrow) caused by an impacted stone. The patient required surgical resection and stone removal.

Courtesy of Jonathan Kruskal, MD.

Graphic 55576 Version 3.0

Mirizzi syndrome seen on endoscopic retrograde cholangiopancreatography (ERCP)



Endoscopic retrograde cholangiopancreatography in a patient with obstructive jaundice and Mirizzi syndrome shows an impacted stone in the distal cystic duct which is obstructing the common hepatic duct by extrinsic compression (arrow).

Courtesy of James B McGee, MD.

Graphic 76550 Version 3.0

Classification of jaundice according to type of bile pigment and mechanism

Unconjugated hyperbilirubinemia
Increased bilirubin production*
Extravascular hemolysis
Extravasation of blood into tissues
Intravascular hemolysis
Dyserythropoiesis
Wilson disease
Impaired hepatic bilirubin uptake
Heart failure
Portosystemic shunts
Some patients with Gilbert syndrome
Certain drugs¶ – Rifampin, probenecid, flavaspadic acid, bunamiodyl
Impaired bilirubin conjugation
Crigler-Najjar syndrome types I and II
Gilbert syndrome
Neonates
Hyperthyroidism
Ethinyl estradiol
Liver diseases – Chronic hepatitis, advanced cirrhosis
Conjugated hyperbilirubinemia
Defect of canalicular organic anion transport
Dubin-Johnson syndrome
Defect of sinusoidal reuptake of conjugated bilirubin
Rotor syndrome

Conjugated hyperbilirubinemia (continued)
Extrahepatic cholestasis (biliary obstruction)
Choledocholithiasis
Intrinsic and extrinsic tumors (eg, cholangiocarcinoma, pancreatic cancer)
Primary sclerosing cholangitis
AIDS cholangiopathy
Acute and chronic pancreatitis
Strictures after invasive procedures
Certain parasitic infections (eg, *Ascaris lumbricoides*, liver flukes)
Intrahepatic cholestasis
Viral hepatitis
Alcohol-associated hepatitis
Non-alcohol-associated steatohepatitis
Chronic hepatitis
Primary biliary cholangitis
Drugs and toxins (eg, alkylated steroids, chlorpromazine, herbal medications [eg, Jamaican bush tea], arsenic)
Sepsis and hypoperfusion states
Infiltrative diseases (eg, amyloidosis, lymphoma, sarcoidosis, tuberculosis)
Total parenteral nutrition
Postoperative cholestasis
Following organ transplantation
Hepatic crisis in sickle cell disease
Pregnancy
End-stage liver disease

AIDS: acquired immunodeficiency syndrome.

* Serum bilirubin concentration is usually less than 4 mg/dL (68 mmol/L) in the absence of underlying liver disease.

¶ The hyperbilirubinemia induced by drugs usually resolves within 48 hours after the drug is discontinued.

Graphic 55607 Version 13.0

Contributor Disclosures

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