

Therapeutic Approach in Grade Iv Mirizzi Syndrome in the Context of Chronic Cholelithiasis: A Rare Complication

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ABSTRACT

Mirizzi syndrome is a rare but significant complication of chronic cholelithiasis characterized by extrinsic compression or erosion of the common hepatic duct caused by impacted stones in the gallbladder infundibulum or cystic duct. The Csendes classification stratifies the syndrome into five grades according to the extent of biliary involvement, from external compression without fistula (Grade I) to extensive cholecystoenteric fistulas (Grade V). Grade IV, the most severe form, involves a wide cholecystobiliary fistula with destruction of more than two-thirds of the diameter of the common hepatic duct, posing significant surgical challenges.

This case report describes a 56-year-old female patient with chronic cholelithiasis presenting with right upper quadrant abdominal pain. Imaging studies initially suggested choledocholithiasis; however, during surgery, Grade IV Mirizzi syndrome with a broad cholecystobiliary fistula and significant bile duct destruction was found. Management included preoperative endoscopic retrograde cholangiopancreatography (ERCP) with stone extraction, followed by laparoscopic cholecystectomy converted to open surgery due to complex biliary anatomy. Reconstruction was performed via Roux-en-Y hepaticojejunostomy, with favorable postoperative evolution. This case highlights the importance of high clinical suspicion, advanced imaging techniques, and surgical expertise for appropriate diagnosis and management of Mirizzi syndrome, especially in its advanced forms, to prevent complications and optimize patient outcomes.

KEYWORDS: Mirizzi syndrome, chronic cholelithiasis, cholecystobiliary fistula, Csendes classification, Roux-en-Y hepaticojejunostomy, choledocholithiasis, biliary complications, surgical diagnosis, surgical treatment, endoscopic retrograde cholangiopancreatography (ERCP).

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INTRODUCTION

Cholelithiasis is one of the most prevalent biliary tract diseases, often asymptomatic or presenting with common complications such as acute cholecystitis or choledocholithiasis. However, in some patients, it may evolve into unusual and complex clinical entities like Mirizzi syndrome. First described by Pablo Luis Mirizzi in 1948, this syndrome is characterized by mechanical obstruction of the common hepatic duct secondary to an impacted stone in the cystic duct or gallbladder infundibulum, resulting in either extrinsic compression or cholecystobiliary fistula formation.

Mirizzi syndrome is a rare complication with an estimated incidence of 0.05% to 4% among patients undergoing cholecystectomy. Its clinical and radiological recognition can be challenging due to symptom overlap with other hepatobiliary diseases such as gallbladder cancer or biliary tumors. The clinical presentation often includes obstructive jaundice, right upper quadrant pain, dark urine, pale stools, and elevated liver enzymes, though findings vary depending on the severity of involvement.¹

The Csendes classification is the most widely used system to categorize Mirizzi syndrome, dividing it into five grades based on the extent of biliary tract damage. Grade IV, the most severe form, involves destruction of more than two-thirds of the common hepatic duct diameter due to a large cholecystobiliary fistula. This advanced grade poses significant surgical challenges, often requiring complex reconstructive procedures like Roux-en-Y hepaticojejunostomy and is associated with higher intraoperative complication risks, especially if the diagnosis is not established preoperatively.^{2,3}

Treatment of Grade IV Mirizzi syndrome demands careful planning and a multidisciplinary approach where

imaging techniques—such as ERCP, magnetic resonance cholangiopancreatography (MRCP), or intraoperative cholangiography—play a crucial role in delineating biliary anatomy and defining the optimal surgical strategy.

This article aims to present a clinical case of Grade IV Mirizzi syndrome in a patient with symptomatic chronic cholelithiasis, describing the therapeutic approach, intraoperative findings, and postoperative outcomes to contribute to the clinical and surgical understanding of this rare and challenging entity.^{4,5,6}

CLINICAL CASE

A 63-year-old female patient with a history of postmenopausal osteoporosis treated with calcium and vitamin D, no prior surgeries or family history of biliary diseases, presented to the emergency department with two days of colicky right upper quadrant abdominal pain rated 7/10 on the visual analog scale, radiating to the right scapular region, without identifiable triggers or relief from self-administered analgesics. She denied fever, nausea, vomiting, dark urine, pale stools, jaundice, changes in bowel habits, or weight loss, and reported no prior similar episodes. Physical examination showed a patient in pain, without pallor or jaundice. The abdomen was distended with moderate subcutaneous fat, soft, with preserved peristalsis and tenderness on deep palpation in the epigastrium and right upper quadrant. Murphy's sign was negative, and there were no signs of peritoneal irritation or palpable hepatomegaly. Vital signs were within normal limits (BP: 120/75 mmHg, HR: 78 bpm, RR: 16 bpm, Temp: 36.7°C, SpO₂: 98%).



Figure 1. Abdominal ultrasound showing a thick-walled gallbladder with multiple gallstones inside, along with dilation of the intrahepatic bile ducts. Additionally, two hyperechoic images of approximately 15 mm are observed in the supraduodenal portion of the common bile duct, compatible with choledocholithiasis.

Abdominal ultrasound revealed a gallbladder with thickened walls containing multiple stones, intrahepatic biliary dilation, and two hyperechoic images approximately 15 mm in the supraduodenal common bile duct compatible with choledocholithiasis.

Preoperative management included ERCP confirming the presence of common bile duct stones, followed by successful endoscopic sphincterotomy and extraction of three 15 mm stones without complications.

Elective laparoscopic cholecystectomy was scheduled but converted to open surgery due to difficult anatomy and distortion of Calot's triangle. Intraoperatively, the gallbladder was enlarged (10 x 6 cm) with thickened edematous walls and multiple stones, some impacted in the infundibulum. A broad cholecystobiliary fistula communicating with the common hepatic duct was identified, consistent with Grade IV Mirizzi syndrome per Csendes classification, showing more than 66% destruction of the bile duct diameter.

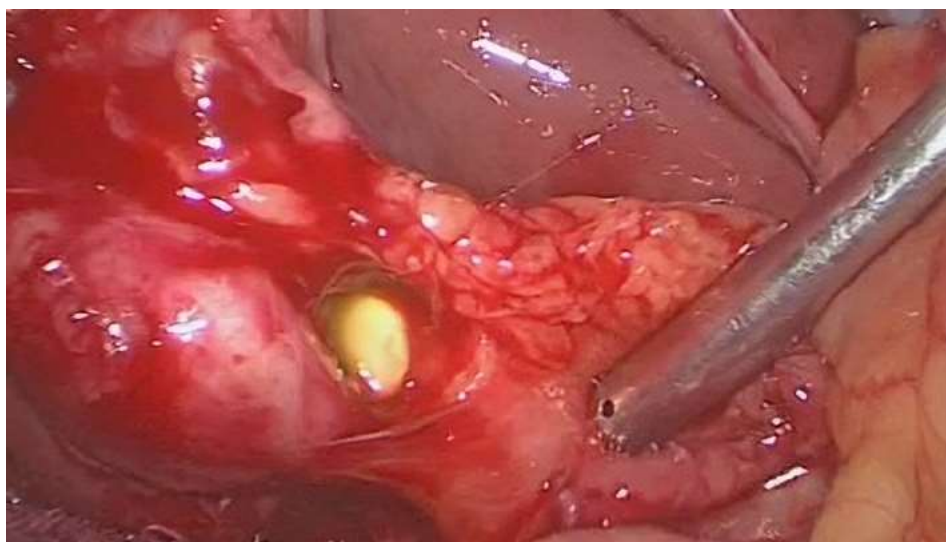


Figure 2. Cholecystobiliary fistula with wide communication to the common hepatic duct, consistent with Mirizzi syndrome grade IV.

A subtotal cholecystectomy was performed, followed by biliodigestive reconstruction using a Roux-en-Y hepaticojejunostomy. Surgery concluded without major complications and with adequate hemostasis.

Postoperative recovery was uneventful; the patient resumed bowel movements by day three, tolerated progressive diet, and had the drain removed on day six. She was discharged on day seven with outpatient follow-up by general surgery and gastroenterology. Subsequent controls showed good clinical and biochemical evolution without evidence of late biliary complications.

DISCUSSION

Mirizzi syndrome is an uncommon complication of chronic cholelithiasis characterized by extrinsic compression or erosion of the main bile duct secondary to an impacted stone in the gallbladder infundibulum or cystic duct. Clinical presentation can mimic other biliary diseases such as choledocholithiasis, cholangitis, or even biliary tumors, complicating preoperative diagnosis.

The Csendes classification divides Mirizzi syndrome into five grades based on bile duct damage:

Grade I: External compression of the common hepatic duct without fistula.

Grade II: Cholecystobiliary fistula involving less than 33% of bile duct diameter.

Grade III: Fistula involving 33–66% of bile duct diameter.

Grade IV: Extensive fistula with destruction of more than 66% of the bile duct diameter.

Grade V: Any of the previous grades plus a cholecystoenteric fistula.

Grade IV, as presented in this case, poses significant surgical challenges due to anatomical distortion, risk of iatrogenic bile duct injury, and the need for complex reconstructive surgery. Our patient presented with typical biliary disease symptoms but no overt cholangitis or jaundice, likely due to partial bile drainage via the fistula.^{7,8} Ultrasound and ERCP initially diagnosed choledocholithiasis; however, intraoperative findings revealed an extensive cholecystobiliary fistula and significant bile duct destruction, confirming Grade IV Mirizzi syndrome.

Literature reports low rates of preoperative diagnosis (10–50%) with MRCP and ERCP being the most useful diagnostic tools, although they may not fully reveal the extent of bile duct destruction. Surgical discovery of a fistula highlights the need to consider this diagnosis in patients with chronic cholelithiasis and recurrent biliary pain episodes. Surgical treatment of Grade IV Mirizzi syndrome is primarily open due to distorted anatomy and injury risk. Roux-en-Y

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hepaticojejunostomy is the preferred reconstruction method to ensure bile drainage and minimize postoperative stricture or leak.^{8,9}

The patient's postoperative course was favorable, reinforcing the importance of clinical suspicion, appropriate imaging, and experienced surgical teams for advanced biliary reconstructions. Timely diagnosis and expert surgical intervention are essential to avoid complications such as persistent fistulas, recurrent cholangitis, or long-term biliary strictures.¹⁰

CONCLUSION

Mirizzi syndrome is a rare but significant complication of chronic cholelithiasis characterized by extrinsic compression or fistula formation between the gallbladder and main bile duct due to impacted stones in the infundibulum or cystic duct. It presents a diagnostic and therapeutic challenge with potential complications from mismanagement.

The Csendes classification is crucial for categorizing Mirizzi syndrome according to biliary involvement, guiding clinical and surgical approaches:

Grade I: External compression without fistula, generally with good prognosis after simple cholecystectomy.

Grades II and III: Cholecystobiliary fistulas involving less than 33% and 33–66% of the bile duct diameter, requiring more careful surgical techniques and sometimes partial reconstruction.

Grade IV: Extensive bile duct destruction over two-thirds of the diameter, necessitating complex biliodigestive reconstruction such as Roux-en-Y hepaticojejunostomy.

Grade V: Presence of cholecystoenteric fistula complicating management further.

This case of Grade IV Mirizzi syndrome underscores the importance of multidisciplinary care, high suspicion in patients with chronic cholelithiasis and biliary symptoms, and the utility of advanced imaging and surgical expertise to achieve favorable outcomes.

Early diagnosis and appropriate surgical management reduce the risk of intraoperative injuries, postoperative complications, and improve quality of life.^{11,12,13}

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