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## MIRIZZI SYNDROME: A SURGICAL CHALLENGE

*Síndrome de Mirizzi: um grande desafio cirúrgico*

Patrícia de Souza **LACERDA**, Manuel Rios **RUIZ**,  
Ana **MELO**, Leonardo Simão **GUIMARÃES**,  
Rubem Alves da **SILVA-JUNIOR**, Gerson Suguiyama **NAKAJIMA**

From the Clínica Cirúrgica, Hospital Universitário Getúlio Vargas, Universidade Federal do Amazonas (Surgical Clinic, Getúlio Vargas Hospital, Federal University of Amazonas), Manaus, AM, Brazil

### Correspondence

Gerson S. Nakajima

E-mail: gnakajima@ufam.edu.br

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### INTRODUCTION

The Mirizzi syndrome consists of the obstruction either of the common hepatic duct or the choledocus, secondary to the extrinsic compression due to the impact of calculus in the cystic duct or in the gallbladder infundibulum<sup>4</sup>. The first description is due to Pablo Mirizzi (1948), when he observed some factors which could cause extra hepatic cholestasis in certain groups of patients carrying cholelithiasis<sup>9</sup>. It generally occurs in female with advanced age. Depending on the degree of involvement of the biliary tract, the patients may be grouped into five distinct groups according to the new rating of the Mirizzi syndrome<sup>1,6</sup>.

The goal of this report is to present a case of a patient carrying type IV, surgically treated through the laparotomy approach.

### CASE REPORT

A 56-year-old woman was admitted into the Service of Surgery of Getúlio Vargas Hospital with history of pain in the right hypochondrium with dorsal irradiation, daily evening fever during three months, and a previous episode of choloria lasting 10 days. The physical examination of admission was normal; laboratory tests demonstrated transaminases changes: glutamic oxalacetic transaminase: 75U/L and glutamic pyruvic transaminase: 62U/L; and the canalicular enzymes: alkaline phosphatase: 1924 U/L, and gama

glutamyl transferase: 884 U/L; the bilirubins were normal. The abdomen ultrasound showed cholelithiasis, bile duct of increased caliber measuring 1.7 cm, showing the "double barreled" with the portal vein, with presence of hyperechoic image measuring 1.1 cm, compatible with calculus; intra-hepatic biliary tract had normal sonographic appearance. The magnetic cholangiography showed cholelithiasis with intra and extra-hepatic biliary tract dilation up to the level of the distal common bile duct, which measured 1.6 cm.

The patient underwent surgical procedure through the laparotomy approach with right subcostal incision. Intraoperatively, there were adhesions of the transverse colon, duodenum and stomach in the gallbladder, which was found to be scleroatrophic and full of calculus. It was decided to perform anterograde cholecystectomy (Torek's). During the procedure, it was observed the presence of fistula between the gallbladder infundibulum and the choledocus, with erosion of its entire anterior wall covering from the implantation of the cystic duct to the proximity of the duodenum, which was classified as type IV Mirizzi. Choledococopy through the fistula was performed with flexible choledocoscope, and a single calculus in the distal choledocus, was identified and removed. The irrigation of the biliary tract with physiological saline solution without elimination of additional calculus, and endside choledocojejunal anastomosis in Roux-en-Y was performed with ligature of the distal choledocus. The peritoneal cavity was drained with latex laminar drain. The diet was released on the second day after surgery, with good acceptance. The drain debit oscillated between 20 and 755 ml, showing bilious secretion until the 10<sup>th</sup> day, when it was removed due to the volume reduction. The patient was discharged on the 12<sup>th</sup> day after surgery.

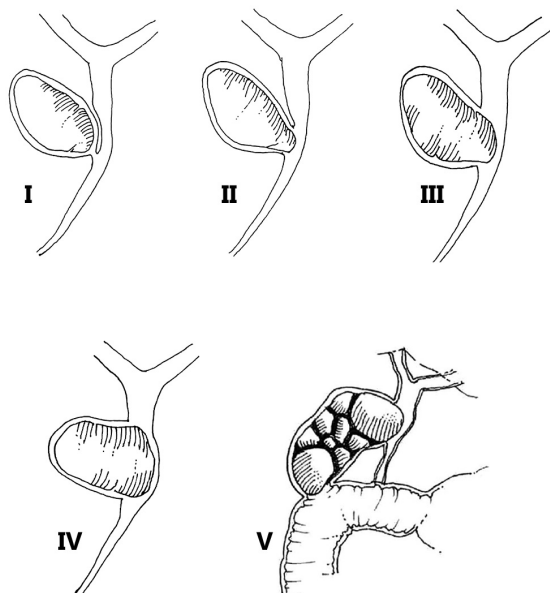
### DISCUSSION

Mirizzi syndrome is a rare complication and it occurs approximately on 0.5 to 4% of the patients carrying cholelithiasis. It's more frequent on women between 21 to 90 years old, probably a reflection of the gallstones preponderance in this group. It is the complication of long standing cholecistolithiasis<sup>5,6</sup>.

The constant compression of the calculus associated to the inflammation of the involved structures may result in fistula between the gallbladder infundibulum or the cystic duct, and the extra hepatic biliary tract. In the cholecystobiliary fistula, the calculus may migrate to the main biliary tract, while in the coloentericystic fistula the patient may show intestinal obstruction called biliary ileus<sup>9</sup>.

The importance of the recognition the Mirizzi syndrome derives from the high risk of lesions of the biliary duct during the surgical procedures. Summing up to this fact is the difficulty for the preoperative diagnosis because there is no specific clinic and laboratory presentation<sup>4,7,10</sup>. The most frequent signs and symptoms are abdominal pain followed by jaundice and cholangitis. Nausea, vomits, choluria, itch, hepatomegaly and, less frequently, acute pancreatitis, gallbladder perforation and weight loss<sup>3,4,5,8</sup>.

The Mirizzi syndrome, which was previously classified into four types, currently the coloentericystic fistula is being included in as complication (type V)<sup>1</sup> (Figure 1). The types are: I) extrinsic compression of the common/choledocus hepatic duct by calculus in the gallbladder infundibulum or cystic duct; II) presence of cholecystoenteric biliary fistula with erosion of the diameter less than 1/3 of the common/choledocus hepatic duct circumference; III) presence of cholecystoenteric biliary fistula with a diameter bigger than 2/3 of the common/choledocus hepatic duct circumference; IV) presence of cholecystoenteric biliary fistula involving the entire common/choledocus hepatic duct circumference; V) any type, plus cholecystoenteric biliary fistula (Va - without biliary ileus, and Vb - with biliary ileus).



**FIGURE 1** - Outline representation of the new classification of Csendes to Mirizzi syndrome<sup>1</sup>

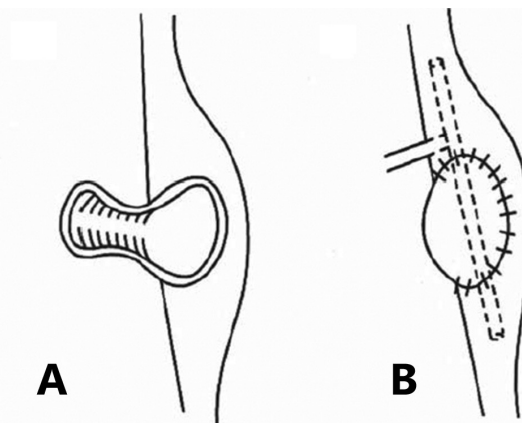
The surgical treatment of the Mirizzi syndrome requires ability and care in the dissection of the biliary tract in order to perform the cholecystectomy, a safe operation of the biliary tract can be avoided and the removal of the calculus so can avoid any iatrogeny in the biliary tract, as in this particular case, where was opted to dissect the biliary tract incompletely through the Torek technique due to the intense inflammatory process<sup>3,5</sup>.

Intraoperatively, perivisceral firm adhesions is found, the gallbladder in most of the cases is scleroatrophic, with or without cholecystoenteric fistula, the Calot's fibrous triangle should arouse the suspicion of this entity. The cholangiography performed by puncture or by Kehr drain as the first procedure is mandatory so that can outline the anatomy of the biliary tract<sup>2,5,11</sup>.

Some authors do not consider laparoscopy as first option due to the intense inflammatory process caused by the disease, being even considered a contraindication to the treatment minimally invasive, but it can be safely performed by experienced surgeons in some cases<sup>1,3,5,6</sup>.

In the absence of cholecystobiliary fistula (Type I), the cholecystectomy and the removal of the biliary calculus constitute the treatment of choice. In the presence of lithiasis of common biliary duct, and when the choledocostomy shows technical difficulties, endoscopic retrograde cholangiopancreatography in the postoperative with the removal of the calculus may be a safe alternative<sup>10</sup>.

In types II and III, the dissection of the cystic duct and the exposure of the Calot triangle may lead to the opening a fistulous orifice in the common biliary duct. In such situation, one of the alternatives is to use technique on which partial cholecystectomy is performed through anterograde via with preservation of the infundibulum, followed by opening of the gallbladder, removal of the calculus of its interior, and choledocoplasty with suture of the fistulous orifice on the remaining wall of the gallbladder. The Kehr drain is introduced into the common hepatic duct over the repair site. The closing of the orifice must be made with no tension and with the stump mucosal of the gallbladder juxtaposed to the duct mucosal. The use of the gallbladder infundibulum to close the orifice of the common hepatic duct is good because it consists of vascular tissue and it has mucosal similar to biliary duct. However, there is a tendency for the formation of fibrosis and stenosis on the suture lines of the biliary duct, even when it is carefully performed<sup>10</sup> (Figure 2).



**FIGURE 2** - Outline representation of a choledocoplasty and Kehr drain placing<sup>7</sup>

If the fistula cannot be primarily corrected with the techniques stated above, the biliodigestive anastomosis can be performed. For the reconstruction of the biliary duct in the Mirizzi syndrome type IV, a choledochal and hepaticojejunal anastomosis in Roux-en-Y is usually necessary as a first act, as in the case of the patient here reported<sup>6,10</sup>. According Mirizzi classification, there is a well defined protocol of surgical treatment for different types: type I – partial cholecystectomy; type II – closing of the fistula with suture or choledocoplasty; type III – choledocoplasty; type IV – biliodigestive anastomosis. The biliodigestive anastomosis has been performed for some of the cases of type II when the erosion of the biliary duct was considered significative<sup>5</sup>.

The organs that can be involved in the cholecystoenteric fistula are the stomach, the duodenum and the colon. The choosing procedure for the cholecystoenteric fistula with biliary ileus is the enterolithotomy and the closing of the fistula, and the one for the fistula without biliary ileus is the closing of the fistula orifice.

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