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CLINICAL CASE

Minimally invasive management of Choledochal Cyst in pediatric age

Manejo mínimamente invasivo de Quiste de Colédoco en edad pediátrica

David Schnettler R.[a], Janitza Norín M.[b], Rafael López D.[b]

[a]Servicio de Cirugía Infantil, Hospital Regional de Talca. Facultad de Medicina, Universidad Católica del Maule. Talca, Chile
[b]Interno, Facultad de Medicina, Universidad Católica del Maule. Talca, Chile.

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What do we know about the subject matter of this study?

Choledochal cysts are a spectrum of congenital extra- and intrahepatic biliary tract anomalies, known for many decades. The etiology is still unclear. Early diagnosis and minimally invasive management allow an excellent prognosis for those patients.

What does this study contribute to what is already known?

We report 3 cases of diagnosis and management of choledochal cysts, using high-resolution imaging techniques such as MRI and minimally invasive laparoscopic approach.

Abstract

The choledochal cyst (also bile duct cyst) is a rare condition. It is important to know its clinical presentation, diagnosis, and treatment alternatives, which allow a resolution with low morbidity. **Objective:** to report the clinical diagnosis together with the laparoscopic techniques for the management of the bile duct cyst. **Clinical Cases:** Case 1: 4-year-old preschooler with history of recurrent abdominal pain. Abdominal ultrasound showed a choledochal cyst. Blood amylase levels 111 IU / L. Other tests were normal. Case 2: 5-year-old preschooler with a 5-days history of abdominal pain, vomiting, and diarrhea. He was admitted due to acute pancreatitis (blood lipase 947 IU / L, blood amylase 217 IU / L). Abdominal CT scan reported a lobulated cystic lesion in the hilum of the liver. Case 3: 3-year-old preschooler with recurrent abdominal pain and a 3-day history of epigastric pain and vomiting. Blood amylase and lipase levels were 248 IU / L and 253 IU / L, respectively, diagnosing acute pancreatitis. Abdominal CT scan showed a finding suggestive of a common bile duct cyst. In all 3 cases, the magnetic resonance cholangiopancreatography reported a type I choledochal cyst. All patients underwent laparoscopic surgery, performing cyst resection, and hepaticoduodenostomy. One case presented pneumobilia without requiring specific management, the other two did not present incidents and all remain asymptomatic in the follow-up period that was longer than one year after surgery. **Conclusions:** In the choledochal cyst, clinical suspicion and timely diagnosis with imaging studies and minimally invasive surgery are important, which allow optimal results in the medium- and long term.

Keywords:
Choledochal Cyst;
Pancreatitis;
Biliary Tract Surgical Procedures;
Obstructive Jaundice

Correspondence:
David Schnettler R.
dschnettlerrod@gmail.com

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Introduction

Choledochal cysts (CC) are single or multiple congenital dilatations of the biliary tree. In the '70s, Todani et al proposed a classification of CC, which is currently maintained with minimal modifications. It should be noted that type I cysts (dilatation of the extrahepatic biliary tract) account for 75 to 85% of the reported cases[1] (Figure 1).

CCs represent less than 1% of benign diseases of the biliary tract[2]. Worldwide, they have an incidence between 1:100,000 and 1:150,000 and is much more frequent in the Asian population, where it is 1:1000. The ratio between men and women is 1:3[3,4].

The etiology of CC is uncertain. The most accepted theory is that of Babbitt, which suggests that an anomalous pancreaticobiliary junction would occur outside the ampulla of Vater. The mixture of pancreatic and bile juice would produce inflammation and subsequent dilatation of the common bile duct[2].

The CCs can produce complications secondary to chronic inflammation, such as ductal stenosis, lithiasis, cholangitis, pancreatitis, ruptures, secondary biliary cirrhosis, and long-term neoplasms[5]. Prenatal diagnosis is possible and can be detected from the second trimester onwards[6] in 20% of cases[7]. In the postnatal period, about 20% of cases present the diagnostic triad of abdominal pain, jaundice, and palpable abdominal mass[5,8]. Most of the patients present with only one of these symptoms and the onset of pancreatitis is frequent. Early diagnosis is important to avoid increased morbidity and mortality[5,6,8].

Abdominal ultrasound usually shows a cystic mass in the hepatic hilum, separated from the gallbladder, which may be single or multiple and involve extra or intrahepatic duct. Depending on the type of cyst, it has up to 97% sensitivity to detect CC and is preferred as a diagnostic method since it is accessible and noninvasive[5]. MR cholangiography is used to plan surgery, since it provides a good visualization of the anatomy, without the risk of cholangitis or pancreatitis that endoscopic retrograde cholangiopancreatography could cause[5,9]. If possible, the abdominal CT scan should be avoided due to the exposure to ionizing radiation[9].

The resolution of these cysts is surgical, with removal and subsequent reconstruction of the biliary tract. For decades, these procedures were open surgical interventions. In the '90s, the laparoscopic technique was initiated[10], which allows performing biliary reconstruction by Roux-en-Y hepaticojejunostomy or by hepatic-duodenal anastomosis (HD)[11].

The objective of this article is to report and discuss the clinical diagnosis and laparoscopic techniques for the management of CC.

Clinical Cases

Retrospective analysis of the last 3 years of cases of patients with CC who underwent surgery at the *Hospital de Talca*. There were three pediatric patients. In all cases, at least one year of postoperative clinical, laboratory, and imaging follow-up was completed.

Case Report 1: 4-year-old preschooler, term newborn, with no morbid history. She was referred to the hospital due to a 2-year history of recurrent abdominal pain, so an abdominal ultrasound was performed which reported a CC. The patient was asymptomatic at the time of consultation, and the study was completed with MR cholangiography, reporting a dilatation of the extrahepatic biliary tract, compatible with type I choledochal cyst. Figure 2 presents the images of abdominal ultrasound and MR cholangiography, which shows the characteristic findings of type I choledochal cyst. Among the laboratory tests, levels of serum amylase (111 IU/L) and alkaline phosphatase (306 IU/L) stood out, the rest of the tests were within normal range.

She underwent laparoscopic resection of the cyst, cholecystectomy, and hepatic-duodenal anastomosis, without incidents. At 6 months of follow-up, abdominal ultrasound showed no biliary tract dilatation and laboratory tests presented no alterations. At 22 months of clinical follow-up, the patient was asymptomatic.

Case Report 2: 5-year-old preschooler, term newborn, with history of chronic constipation and recurrent obstructive lung disease, consulted due to 5-day history of colicky abdominal pain, which progressively increased, associated with vomiting and diarrhea, without fever. Physical examination showed generalized abdominal pain, with no other findings. Based on the clinical and laboratory findings (lipase 947 IU/L, amylase 217 IU/L, total bilirubin 1.27 mg/dL, direct bilirubin 0.95 mg/dL, transaminase AST/GOT 303.5 IU/L and ALT/GPT 368.5 IU/L), acute pancreatitis was diagnosed, thus the patient was hospitalized in the Critical Patient Unit. A CT scan of the abdomen and pelvis was requested, which reported a lobulated cystic lesion, affecting the hepatoduodenal ligament and the head of the pancreas. MR cholangiography was requested, which reported a fusiform cystic dilatation of the extrahepatic biliary tract, with 3.3 cm in diameter in the common hepatic duct and distal common bile duct (type I cyst), as well as cholelithiasis. The surgical resection was planned after the resolution of acute pancreatitis.

The patient underwent laparoscopic resection of the cyst, cholecystectomy, and hepatic-duodenal anastomosis, without incidents. One month after surgery, an abdominal ultrasound was performed, which reported possible intrahepatic lithiasis, so a new MR cholangiography was performed, which concluded that the

image corresponded to pneumobilia, without lithiasis nor stenosis, and did not require specific management. Control laboratory tests showed no alterations. In the clinical follow-up 18 months after surgery, the patient has remained asymptomatic.

Case Report 3: 3-year-old preschooler, term newborn, with a 2-year history of recurrent abdominal pain associated with vomiting in pain crises, with no other morbid history. He was referred due to 3-day history of epigastric pain, associated with vomiting refractory to medical management. On physical examination, he had a mass tender on palpation, deep in the epigastrium, with no other findings. Based on clinical and laboratory tests (amylase 248 IU/L, lipase 253 IU/L, other tests within normal range), acute pancreatitis was diagnosed. A CT scan of the abdomen and pelvis was requested, which reported dilatation of the common bile duct, with fluid content, suggestive of CC, without other alterations. MR cholangiography was performed, which reported cystic dilatation of the common bile duct throughout its length (type I), with stones and biliary sludge, without other alterations.

After the resolution of his pancreatitis, he underwent laparoscopic resection of the cyst, cholecystectomy, and hepatic-duodenal anastomosis, all without incidents. Control abdominal ultrasound one month after surgery reported no pathological findings, and laboratory tests did not present alterations. In the

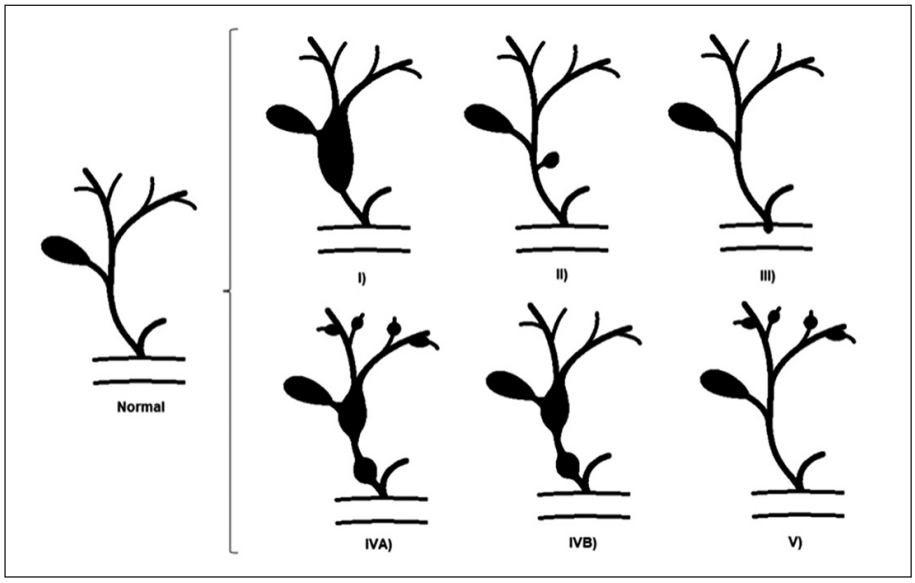

Figure 1. Types of common bile duct cyst, according to location in bile ducts. I) Without intrahepatic component, subdivided into Ia) Spherical, Ib) Segmented and Ic) Fusiform. II) Diverticular dilation that is not limited to the common bile duct. III) Choledocecele, dilation in the wall of the duodenum. IV) Multiple cyst, subdivided into Iva) Intrahepatic and extrahepatic, IVb) Only extrahepatic. V) Caroli's disease, single or multiple dilation of the intrahepatic bile duct[1].

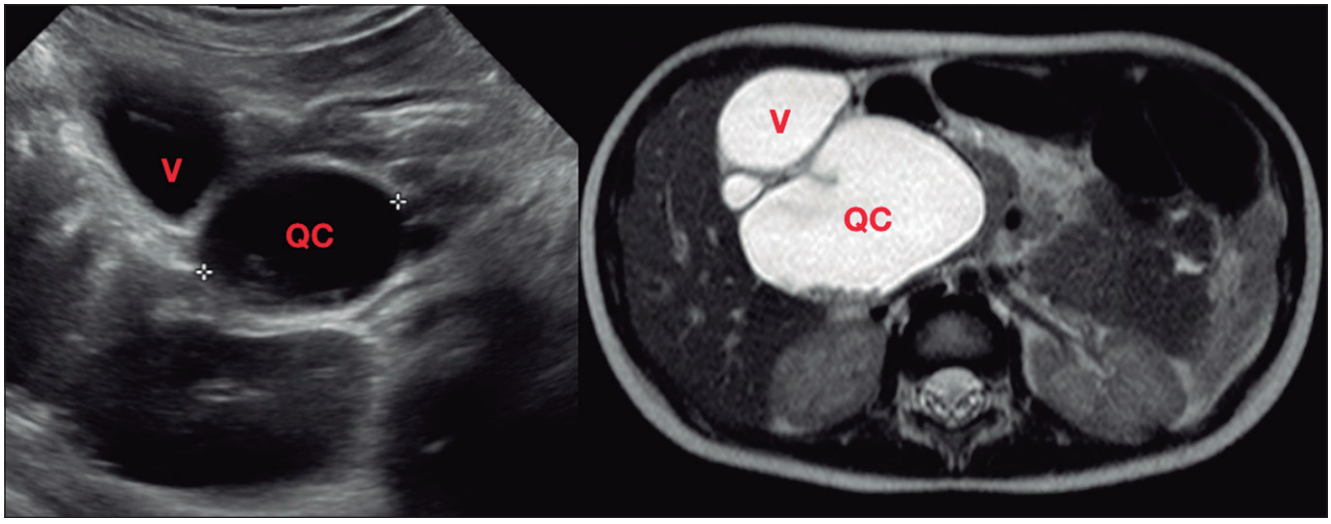

Figure 2. Abdominal ultrasound and cholangio MRI, showing dilation of cysts in the adjacent extrahepatic bile ducts and gallbladder. CC: Coledocal cyst, G: Gallbladder, MRI: Magnetic Resonance Imaging.

clinical follow-up 14 months after surgery, the child has remained asymptomatic.

Regarding the surgical technique, the three cases underwent surgery using 3 mm laparoscopic instruments, 5 mm end effectors at a 30-degree angle, and 12 mmHg-pressured pneumoperitoneum. Intraoperative cholangiography through the gallbladder and liver biopsy were performed. The resection was performed with electrocautery, the hepatic-duodenal anastomosis was performed with absorbable sutures 5-0 with simple running technique and intracorporeal knot-tying, and peri-anastomosis drainage was placed, which was removed after 5 days (figure 3). The average surgery time was 248 minutes. There were no intraoperative or postoperative complications and no conversion to open surgery was required (Table 1).

Discussion

The CC is an infrequent and not very symptomatic pathology in its onset, so it is relevant to analyze the diagnostic process. It is important to maintain a high index of suspicion in the presence of nonspecific digestive manifestations, abdominal pain, jaundice, or pancreatitis. Abdominal ultrasound is a good diagnostic test[5] and MR cholangiography is very useful in determining the anatomy[5] and type of cyst identified[9].

In the cases presented, 2 of 3 had clinical symptoms suggestive of acute pancreatitis, without the presence of cholangitis or clinically significant jaundice. In the third case, the manifestation was recurrent abdominal pain, with incidental finding in abdominal ultrasound. In this series, there were no cases of prenatal diagnosis, nor with the classic triad of CC (jaundice, palpable mass, and abdominal pain)[5,8], and abdominal pain was a constant finding[12,13].

Surgical treatment is the removal of the cyst and subsequent biliodigestive anastomosis, with the main objective of restoring the physiological flow of bile, thus preventing the occurrence of cholangitis, leaks, biliary stenosis, or other medium and long-term complications, such as cholangiocarcinoma[3,11,12].

When comparing open versus laparoscopic surgery, the latter presents benefits such as better cosmetic results, shorter hospital stay, shorter recovery time of intestinal function, less need for intraoperative transfusion, lower risk of intestinal obstruction due to adhesions, and incisional hernias[14,15]. It does not show significant differences regarding pancreatitis or postoperative complications[14,15]. Regarding the evidence, bile leak and abdominal bleeding are controversial[14,15], but in series reporting greater surgical experience of the teams, and lesser blood loss in laparoscopic surgery compared with open surgery[15].

Regarding the type of biliodigestive anastomosis to be performed after resection of the cyst, for decades, Roux-en-Y hepaticojejunostomy has been performed; however, in recent years, several clinical series have been published a hepatic-duodenal (HD) anastomosis[11,16,17,18]. The latter is an expeditious technique that allows endoscopic access to the anastomosis in the case of stenosis or lithiasis, which is very difficult or impossible to perform endoscopically in hepaticojejunostomy[16,17].

Several publications compare both techniques, showing that HD type anastomosis does not present significant differences in terms of postoperative bile

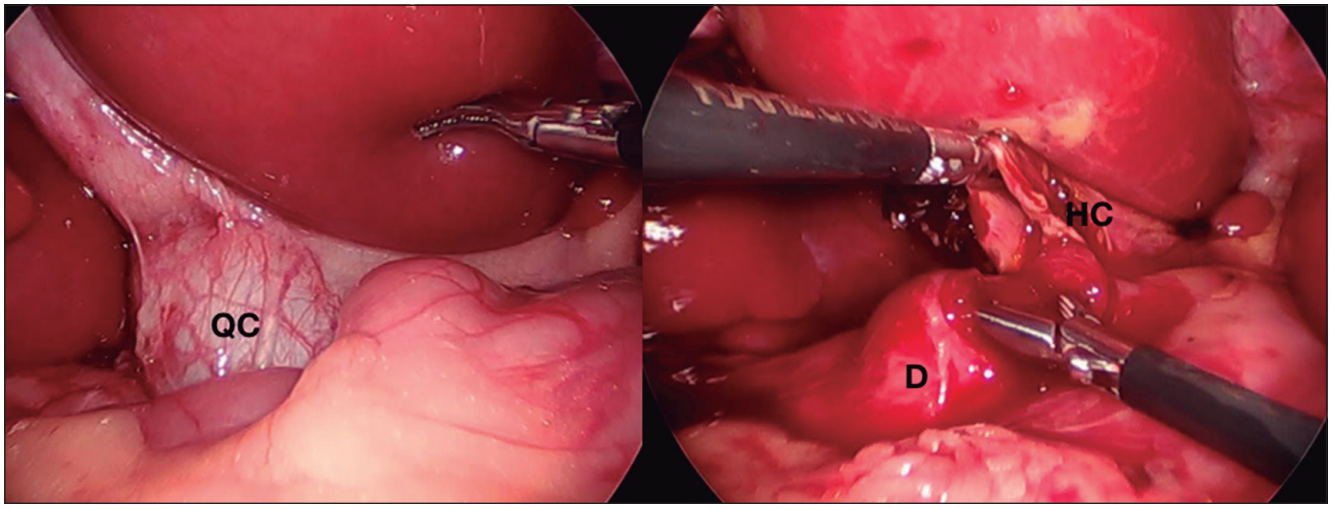

Figure 3. Laparoscopic vision of Choledocal cysts to left and anastomosis hepatic-duodenal to rigth. CC: Coledocal cyst, HC: Hepatic Common Duct, D: Duodenum.

Table 1. Surgery's characteristics

Case	OT	HT	Surgery	Complication
1	228	9	Laparoscopic cystectomy plus HD anastomosis and cholecystectomy	No
2	258	7	Laparoscopic cystectomy plus HD anastomosis and cholecystectomy	No
3	259	7	Laparoscopic cystectomy plus HD anastomosis and cholecystectomy	No

OT: Operative time in minutes, HT: Hospitalization time in days, HD: Hepato-Duodenal

leaks, cholangitis, anastomotic stenosis, adhesive obstruction, and reintervention[11,16,18] and it does present a shorter hospital stay, shorter operative time, less bleeding, and less probability of conversion to open surgery[11,16]. On the other hand, there would be a greater number of patients with duodeno-gastric reflux or gastropathy in those who underwent HD anastomosis[7,16,19], requiring in some series second surgical interventions years later in the most affected cases[19].

Some theories regarding reflux propose that the patient's growth and the stretching of the anastomosis towards the upper part of the duodenum would cause reflux[16,19] and that it could be due to the proximity of the anastomosis to the pylorus[16]. This could be prevented by performing the HD anastomosis as distal as possible in the second portion of the duodenum[17], with about 3 cm distance from the pylorus, which could avoid reflux[16,18,20], reducing the possibility that gastric duodenal reflux could generate carcinogenesis in the long term[16].

All the cases presented underwent HD anastomosis laparoscopically, with intraoperative times similar to other clinical series[16,18], without the need for conversion to laparotomy or other incidents. There were no complications detected in more than one year of follow-up, except for the finding of asymptomatic pneumobilia in one of the cases, which did not require specific management.

Conclusions

In children, the diagnosis of CC should be considered in cases of prenatal cystic mass, and postnatally in cases of recurrent abdominal pain with no other apparent cause, jaundice, abdominal mass, or pancreatitis[6,21]. Definitive treatment is surgical, preferably laparoscopic, by resection of the cyst and biliary-digestive anastomosis[15,18]. The advantages of the laparoscopic approach over the open one are shorter recovery time and lower risk of complications[14,15]. Long-term studies are still needed to define which laparoscopic bilio-digestive anastomosis technique will have better results[11,16,18].

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

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