

## ◆ MANUSCRITO ORIGINAL

# Choledochal cyst in pediatric patients: a 10 years single institution experience

Micaela Germani, Daniel Liberto, Gastón Elmo, Pablo Lobos, Eduardo Ruiz

Department of Pediatric Surgery, Italian Hospital of Buenos Aires, Buenos Aires, Argentina.

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### Summary

**Background.** Choledochal cysts (CCs) are rare congenital dilatations of the biliar tree. The incidence is 1:150.000. A correct diagnosis and surgical treatment are important because of the long term risks of infection and neoplasia. We report our experience with CCs in children. **Patients and methods.** Patients with CC disease, who were seen at the Hospital Italiano de Buenos Aires between 1999 and 2009, were identified retrospectively from our prospectively acquired hepato-pancreatico-biliary database. Prenatal, clinical, surgical and pathological records were analysed. **Results.** We included 12 children (10 girls and 2 boys), all of them affected by a cystic dilatation type I of Todani classification. Between the 4 patients with antenatal diagnosis only one presented symptoms before surgery, expressed in acolic feces. Between the 8 patients with postnatal diagnosis, the most common clinical presentation was abdominal pain, occurring in all patients. Ultrasound was used for the diagnosis of all patients. Mean age at surgery was 74 months. All patients had intraoperative cholangiograms to definitively confirm the diagnosis. Five patients underwent open surgery and 7 laparoscopic approach. From patients with a laparoscopic approach, 1 had to be converted to open surgery due to intraoperative complications and 2 underwent an initial surgery for a cholecystectomy and a CC cyst excision. The complications observed were: anastomotic biliary fistula 1, residual lithiasis in the pancreatic portion of the duct 1 and anastomotic stenosis 1. The last 2 cases required reoperation. **Conclusions.** Once CCs are diagnosed, careful treatment decisions need to be taken because both intra hepatic and extra hepatic CCs, if untreated, have an increased risk of cancer. After a careful study of the cyst's characteristics, we recommend a laparoscopic approach when possible, as the best choice,

after 3 months of life to increase the success of the treatment according to the size of the anatomic structure. We recommend early treatment, before 3 months of life, only in case of severe symptoms.

**Key words.** Choledochal cyst, pediatric, laparoscopic approach

### Quiste de colédoco en niños: la experiencia de 10 años en un centro único

#### Resumen

**Antecedentes.** El quiste del colédoco (QC) es una patología muy poco frecuente que está representada por una dilatación congénita de las vías biliares principales. El diagnóstico precoz y correcto, y un buen tratamiento quirúrgico, son importantes para evitar los riesgos de una transformación maligna. **Métodos.** En este trabajo hicimos una revisión de la casuística de los pacientes pediátricos tratados por esta patología en nuestro centro durante los últimos 10 años (1999-2009). Fueron considerados todos los datos demográficos para cada paciente, además del diagnóstico prenatal, el tratamiento y las patologías relacionadas. **Resultados.** Tratamos 12 niños (10 mujeres y 2 varones) con diagnóstico de QC, todos tipo I según Todani. Cuatro pacientes tuvieron diagnóstico prenatal y entre éstos, solo uno presentó acolia fécal al nacimiento. Todos los casos de diagnóstico postnatal presentaron dolor abdominal. A todos los pacientes les hicimos una ecografía abdominal inicial. La edad media en el momento de la cirugía fue de 74 meses. Realizamos una colangiografía intraoperatoria para el diagnóstico definitivo en todos los pacientes. La cirugía fue abierta en 5 pacientes y laparoscópica en 7. Tres pacientes tuvieron complicaciones luego de la cirugía laparoscópica. **Conclusión.** Después del diagnóstico de QC por el estudio de la vía biliar recomendamos el tratamiento laparoscópico en cuanto sea posible, después de los 3

Correspondence: Micaela Germani  
E-mail: micaela\_germani@yahoo.it

*meses de vida en los pacientes asintomáticos o con sintomatología leve.*

**Palabras claves.** *Quiste del colédoco, pediatría, laparoscopia.*

By definition, choledochal cyst is a dilatation of the biliary tree. It is thought to be congenital and is a very rare entity in Western countries. Most of the reported cases in the world come from Asia, with an incidence of 1:1000. About 2/3 of cases were reported in Japan. In recent years, cases of choledochal cyst are reported increasingly in China.<sup>1</sup> The reason of this geographic distribution, the etiology, the adequate classification system, the ideal diagnostic, the therapeutic modalities and the natural course remain unknown. Disease patterns across various ages may help in elucidating the pathophysiology and natural course of biliar cystic disease. The largest series of patients with choledochal cyst disease outside of Asia comes from Vancouver, British Columbia, where 70 patients treated between 1971 and 2003, both pediatrics and adults, were studied.<sup>2</sup> Recognition and management of choledochal cyst disease is important because of the risk of developing cholangiocarcinoma. Based on this risk, complete excision of the cyst is considered the standard of care.<sup>2</sup> To further characterize choledochal cyst disease in a predominantly Western population, we performed a retrospective analysis of pediatric patients seen at the Italian Hospital of Buenos Aires from January 1999 to January 2009.

### Materials and Methods

Patients affected by choledochal cyst disease who were seen at the Italian Hospital of Buenos Aires between 1999 and 2009 were retrospectively identified from our prospectively prepared hepato-pancreaticobiliary database. For patients who underwent operative management we evaluated demographic data, medical comorbidities, antenatal diagnosis, presenting symptoms, diagnostic modalities used, laboratory data (pre and post surgery treatment), operative outcomes, surgical strategy used, complications, pathologic findings in choledochal cyst and liver biopsy, and long-term survival after intervention.

Choledochal cyst was defined using the Todani modified Alonso-Lej classification system. Preoperative morbidity and mortality were also evaluated.

Preoperative complications were defined as occurring within 30 days of operation. Long-term follow-up was based on medical record reviews and phone calls. Complete survival information was available for all patients included in this study.

### Results

Table 1 summarizes part of our results. Twelve paediatric patients, with a mean age of 4.9 years (range 1 day to 8 years), were diagnosed as having choledochal cyst. Five of these patients (41%) were less than 1 year old. Ten patients were female (83%) and two male. Antenatal diagnosis was done in 4 patients (33%). According to Todani classification, all patients had type I cysts. Only 1 of the 4 patients with antenatal diagnosis presented symptoms before surgery, expressed as acolic feces. Between the 8 patients with postnatal diagnosis, the most common clinical presentation was chronic abdominal pain occurring in all patients without amilase increase. Abdominal pain was associated with nausea and vomiting in 6 patients (75%), jaundice in 2 (25%), anorexia in 1 (12.5%), and weight loss in 1. None of these patients presented pruritus. None of the 12 patients presented the classic triad of pain, jaundice and palpable mass. Related comorbid illness included 2 patients presenting pancreatitis and biliary lithiasis. Ultrasound (US) was used in the diagnosis of all pediatric patients, most of all for the differential diagnosis of biliary atresia. The diagnosis was confirmed by a second US examination in

**Table 1.** *Patients with choledochal cyst (n = 12).*

	n	%
<b>Sex</b>		
Female	10	83
Male	2	17
<b>Symptoms</b>		
Abdominal pain	12	100
Gastric symptoms	6	50
Jaundice	2	17
Loss of weight	1	8
Pruritus	0	0
Abdominal mass	0	0
<b>Diagnosis and treatment</b>		
Antenatal diagnosis	4	33
Associated malformations	0	0
Open surgery	5	42
Laparoscopic surgery	7	58
Complications	3	25
Choledochal cyst histology	12	100

the 4 cases with antenatal diagnosis. Magnetic resonance imaging (MRI) was performed in 3 patients, computerized tomography (CT) scan in 2, and endoscopic retrograde cholangiopancreatogram (ERCP) in 1. All patients underwent a blood laboratory exams before surgery and 1 week after it. Mean age at surgery was 74 months. All the 12 patients had intraoperative cholangiograms to definitively confirm the diagnosis. Cyst excision, cholecystectomy and Roux-in-Y hepatic jejunostomy (termino-terminal in all cases) were performed in all pediatric patients. Five patients (42%) underwent direct open surgery and 7 (58%) laparoscopic approach. One of these cases had to be converted to open surgery for intraoperative complications and 1 underwent an initial surgery for cholecystectomy and a second one for choledochal cyst excision. Duration of surgery ranged between 120 and 240 minutes for open surgery, with a mean surgical time of 180 minutes, and between 180 and 300 minutes for laparoscopic approach, with a mean surgical time of 240 minutes. In all patients a postoperative abdominal drain was placed for a mean period of 5.25 days. Early post-surgery complications (within the first week) occurred in 1 patient who presented a biliary fistula and the abdominal drain had to be maintained for two weeks. Late post-surgery complications (within three months) occurred in 2 patients who underwent a second open surgery. The first one underwent a re-anastomosis operation due to a hepatic-jejunostomy stenosis. The second one underwent a duodenectomy and excision of intra-pancreatic cyst due to a recurrent pancreatitis caused by lithiasis in the remnant distal choledochal tract. Eleven patients (91%) received metronidazole as postoperative antibiotic, associated with gentamicin in 8 (72%), amikacin in 1, ceftriaxon in 1, and ceftriaxon and amikacin in 1. One patient received gentamicin and cephalexin. After surgery, total parenteral nutrition was administered during a mean period of six days in 4 patients and the remaining cases received enteral nutrition during a mean period of three days.

We evaluated pre- and post-surgery laboratory data: total bilirubin, partial bilirubin, alkaline Phosphatase (AP), alanine aminotransferase (ALT) and aspartate aminotransferase (AST). The mean total bilirubin value was 2.8 mg per dL (normal value 1.4 mg per dL) in the preoperative exam and 0.86 mg per dL in the postoperative exam. The

mean partial bilirubin value was 1.59 mg per dL (normal value 0.4 mg per dL) in the preoperative exam and 0.37 mg in the postoperative exam. The mean PA value was 354.8 IU per L (normal value 74 to 327 IU per L) in the preoperative exam and 207.9 IU per L in the postoperative exam. The mean ALT value was 116 IU per L (normal value 10 to 40 IU per L) in the preoperative exam and 67 IU per L in the postoperative exam. The mean AST value was 157 IU per L (normal value 10 to 42 IU per L) in the preoperative exam and 77 IU per L in the postoperative exam.

The histological examination confirmed the diagnosis of choledochal cyst in the 12 cases. In only 1 case the liver histology showed fibrosis and this patient was posteriorly followed-up by pediatricians.

Length of hospital stay widely varied between 4 and 14 days, with a mean stay of 7.5 days. After surgery, we controlled all patients with clinical evaluation, ultrasound examination and laboratory exams, one week after discharge, once a month during the first three months, twice a year during the first two years, and once a year after this period. Long-term follow-up compliance was good and all patients had a control during the three last months. None of them presented symptoms related to the choledochal cyst and laboratory and US results were normal.

## Discussion

Choledochal cyst is an exceeding rare congenital benign disease of the biliary tree, accounting for approximately 1% of all benign biliar diseases,<sup>3,4</sup> with a great Asian preponderance.<sup>4,5</sup> Familial cases are extremely rare.<sup>6</sup> In this study we report a series of choledochal cysts in paediatric patients that summarizes the most frequent clinical presentations, diagnosis ways and treatment of this rare pathology. The etiology of biliary cysts remains uncertain.<sup>7</sup> The most generally accepted theory is that they are caused by congenital abnormalities in the pancreatic-biliary junction, present in 39% to 96% of patients with choledochal cyst, according to several authors.<sup>7-9</sup> The junction of the common bile duct with the pancreatic duct outside the duodenal wall, forming a long common channel, would favour reflux of pancreatic enzymes into the biliary tract with damage and dilatation of the bile duct wall. However, in some choledochal cyst these particular features are not found.<sup>5,9</sup> A female 4:1 pre-

ponderance of biliary cystic disease is reported in the literature and this gender pattern is echoed in this study but with a higher ratio (5:1).

Choledochal cysts are usually diagnosed during childhood, but 20% of cases are identified during adulthood.<sup>5,7,10</sup> The diagnosis of choledochal cysts by prenatal US allows a prompt intervention in the neonate. The common bile duct can be identified at 15 to 16 weeks of gestation and the median gestational age at the initial diagnosis is 20 weeks. Fetal choledochal cyst usually appears as a fluid filled, anechoic mass in the region of the *porta hepatis*. The demonstration of a continuity with the gallbladder or hepatic ducts, or associated dilatations of intra- and extra-hepatic ducts further aids to the diagnosis. The prenatal differential diagnosis of choledochal cyst includes simple hepatic cyst, biliary atresia, ovarian, omental or mesenteric cysts, duodenal or gallbladder duplications, adrenal cyst, renal cyst, dilated loops of the bowel, hydronephrotic renal pelvis and *situs inversus*.<sup>11-13</sup> In our series US was the imaging modality used in the initial postnatal diagnosis of choledochal cysts. In order to better classify the cyst some of our patients successively underwent more specific examinations as ERCP, TC scan or MRI.<sup>1</sup>

Todani et al described a classification system of these cysts into six discrete types, which is still in use.<sup>14</sup> The most common types are I and IV A and all our patients presented type I cysts.<sup>6,14,15</sup> Type I and IV A cysts are thought to be due to the abnormal pancreatic-biliary duct junction already described.<sup>8,9</sup> The most common symptoms in our series were non-specific upper abdominal pain, nausea and vomiting, accounting for the fact that bile, pancreatic juice and bile stasis lead to chronic inflammation and stone and stricture formation. Afterwards recurrent cholangitis, hepatic abscesses and pancreatitis appear, causing significant pain. In our series, comparing newborns with infants, the last group had a significant higher number of preoperative morbidities, abnormal elevation of serum aminotransferases and liver fibrosis. These features suggest that choledochal cyst during infancy is a progressive disease. A therapeutic plan for newborns and infants with choledochal cyst is important because of the potential complications later in the life, including biliar cirrhosis and portal hypertension.<sup>8</sup> Furthermore, patients with choledochal cyst have a substantially higher risk of developing

cholangiocarcinoma when compared with the general population. This risk may be due to sustained reflux of pancreatic enzymes and stagnation of bile salts, causing chronic inflammation of the cystic mucosa and subsequent malignant degeneration.<sup>16,17</sup> If this occurs, survival is very poor and most patients die within 2 years. The optimal timing for intervention in neonates has not yet been established, although it is universally accepted that complete extra-hepatic biliary excision, including the gallbladder, is the treatment of choice. It has been suggested that progressive intra hepatic ductal dilatation, cyst enlargement and deterioration of liver function are harbingers of obstruction and/or cholangitis, prompting early surgical intervention.<sup>18</sup> Although prenatal internal drainage procedures without cyst excision have been a commonplace in the past, it has been shown that they predispose to malignancy. The reasons are not entirely clear, but proposed mechanisms include biliary stasis, biliary lithiasis infections, recurrent cholangitis, pancreatitis and conversion of bile salts to carcinogenic substances by chronic infections.<sup>18</sup> In the past, the accepted surgical treatment of choledochal cyst was drainage into the adjacent duodenum or stomach. Because of recurrent episodes of cholangitis, surgeons started draining the cyst with a Roux-in-Y jejunal loop.<sup>18</sup> A lot of reports have demonstrated that internal drainage procedure without resection carries a high morbidity rate and often requires a subsequent reparative operation.<sup>17</sup> Todani analyzed the problem of carcinoma arising from retained cysts with internal drainage procedures and suggested that enteric drainage tends to create a *cul-desac* in the choledochal cyst and activate pancreatic juice.<sup>17,19</sup> The inflammation of the bile duct wall is accelerated as a consequence, probably resulting in carcinoma development because of the long-standing irritation of the biliary epithelium. Kobayashi points out that the incidence of bile duct carcinoma is still high even after excision of extra-hepatic bile ducts in patients with choledochal dilatation.<sup>19,20</sup> For these patients, a careful long-term follow-up is necessary, especially after operation. With the increasing awareness of lifetime risk of cholangiocarcinoma, the standard of care for patients with choledochal cysts has become the complete cystectomy and the hepatic jejunostomy through an intestinal Roux-in-Y loop, it is essential to know the anatomical details and variants of extra-

and intra-hepatic biliary tree as well as adjacent hepatic arterial and portal venous structures, to obtain a better approach to this kind of surgery. An adequate exposure, allowing full visualization, is necessary for a good biliary-enteric anastomosis. In this series, all patients underwent complete excision of the cyst and reconstruction of the anatomic continuity using a Roux-in-Y hepatic jejunostomy.

There are reports in the literature showing that liver fibrosis progresses faster in the newborn cases.<sup>21</sup> Only one patient of our series showed liver fibrosis. Laparoscopic approach, first described in 1995 by Farello et al in a 6-year-old child with a choledochal cyst, nowadays has become a very popular technique for the treatment of this pathology because it is minimally invasive, causes less pain and recovery after surgery is shorter.<sup>22-25</sup> The potential disadvantages of laparoscopic surgery for complex biliary surgery are the loss of direct hepatic feedback, the translation of 3-dimensional structures into a 2-dimensional video image, and the limitation of movements of the laparoscopic instruments compared to human hands. The potential advantages include decreased pain, improved cosmetics and improved visualization with higher magnification through the endoscope.<sup>26,27</sup> Progressive improvements and instrumentation in the last 15 years have led to the development of 3 mm short laparoscopes specially designed for its use in the pediatric population. If the dissection is kept as closely as possible to the wall without isolating the portal vein and the hepatic artery, the procedure becomes safe and fast. As in the open procedure, the curved needle 5-0 PDS suture is helpful for the anastomosis between the hepatic duct and the jejunal limb. Postoperative complication leading to a second surgery occurred in two patients. In the patient reoperated for a stenosis of the bilio-digestive anastomosis, the complication was probably related to a very extensive cyst resection without maintaining a rim of dilated hepatic duct to simplify the anastomosis. The second complicated case underwent a reoperation due to lithiasis of the remnant distal choledochal intra-pancreatic tract. These complications occurred in the beginning of our laparoscopic experience. Learning curve seems to be very important in this complex surgery, especially in small children.

Patients with choledochal cyst require a close and regular follow-up, mainly aimed to detect po-

tential cholangiocarcinoma early.<sup>17</sup> The development of cancer after excision occurs in less than 1% of patients and most of these cases occurs in the setting of an incomplete resection.<sup>28</sup> We controlled all patients with periodical visits to the hospital because follow-up should last for life. Complete cyst resection substantially reduces this risk but does not exclude it completely and there are several reports of cholangiocarcinoma years after cyst excision.

In conclusion, reviewing this series we found that the majority of patients were female and presented type I cysts according to the literature.<sup>5,6,29</sup> In opposite to the literature we did not find the classic symptomatic triade. The most common presenting symptom was intermittent jaundice in children younger than three months old and abdominal pain associated with nausea and vomiting in children older than one year old. Once choledochal cyst has been diagnosed, careful therapeutic decisions need to be taken because if it remain untreated the risk of cancer is increased.<sup>16,17,19,28</sup> The surgical strategy should be selected based on the type of cyst: cystectomy with Roux-in-Y hepatic jejunostomy in type I, ERCP in type III and initial medical treatment in Caroli's disease in type IV A, although hepatic lobectomy and liver transplant could be required. At least extra-hepatic biliary tract cysts should be resected when possible.<sup>23-25,30</sup> The optimal surgical timing in infants, especially in the asymptomatic newborns, is still unknown due to the technical difficulty of surgery at an early age, especially in the laparoscopic approach, and the possible anastomotic complications. Early surgical intervention has been suggested in patients with cyst of rapid expanding size, deterioration of liver function and obstructive jaundice.<sup>22,23</sup> After a careful study of the cyst's characteristics, we recommend laparoscopic approach when possible, as the best choice after three months of life to increase the success of the treatment according to the size of anatomic structures. We recommend early treatment, before three months of life, only in severe symptomatic patients. There is a clear rationale for the laparoscopic approach to choledochal cyst because it is well known that the majority of these patients will enjoy a good quality of life once the abnormal dilated duct is excised. Under these circumstances, the excellent cosmetic results are welcomed in comparison with the large incisions

usually required for conventional operation. These benefits, added to less pain, prompt oral feeding and rapid hospital discharge, make us recommend the laparoscopic approach. Long term follow up is mandatory in patients with choledochal cyst to detect complications, such as malignant degeneration, early, except in type III choledochal cysts that very rarely transform to cholangiocarcinoma.<sup>21</sup>

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