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ORIGINAL ARTICLE

Choledochal cyst: a 10-year experience

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Abstract

Objective: to assess the outcome of surgical treatment of choledochal cyst in patients within a 10-year period.

Methods: retrospective review of medical records of 18 patients submitted to surgical treatment of choledochal cyst.

Results: eighteen patients aged 20 days to 13 years, 15 (83%) female and 3 (17%) male. Initial symptoms: 15 (83%) patients presented with jaundice, 11 (61%) with dark urine, 10 (55%) with acholic feces, 9 (50%) with abdominal pain, and 2 (11%) with abdominal mass. Fourteen (77%) patients were diagnosed by sonography of the abdomen. Endoscopic retrograde cholangiopancreatography was carried out in four patients, and indicated an abnormal common hepatic duct in three of them. Nine patients presented type I, one presented type II, seven presented type IV, and one presented type V cysts. All patients, with the exception of the patient with type V cyst, were submitted to cyst excision with Roux-en-Y hepaticojejunostomy. Two (11%) postoperative deaths were recorded due to complications related to the deterioration of hepatic function. Out of the 15 patients who were submitted to long-term follow-up, two (14%) presented chronic pancreatitis, and 13 (86%) were free of symptoms, without any evidence of late complications.

J Pediatr (Rio J) 2000; 76(2):143-8: choledochal cyst, anastomosis, Roux-en-Y, treatment outcome, cholestasis.

Introduction

Choledochal cyst is a rare abnormality which is usually diagnosed in pediatric female patients. The estimate of incidence of choledochal cyst in the Western-hemisphere population is of 1:100,000-150,000 live births.[1]

The etiology of congenital dilatation of the common bile duct has not been completely defined yet. Different studies have indicated that it is a congenital abnormality, whereas others have indicated that it is an acquired process. The most widely accepted explanation as to the origin of choledochal cyst is that it may be related to chronic reflux of pancreatic juice into the bile duct, secondary to an abnormal connection between the pancreatic duct (Wirsung's duct) and the common bile duct. Different authors have observed that up to 80% of the patients with choledochal cyst have presented that abnormality.[2-4]

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The importance of early diagnosis of choledochal cyst lies in the complications inherent to its development, including possibility of canceration, risk of biliary atresia, cholangitis, spontaneous or traumatic rupture of the cyst, progressive biliary cirrhosis, and portal hypertension.[6]

In 1959, Alonso-Lej et al. presented the first classification system for choledochal cyst.[6] Later, Todani et al. developed a new system including patients with intrahepatic dilatation.[7] The latter is the most frequently used classification system.

During the last few decades, the most widely accepted surgical treatment has been cyst excision with Roux-en-Y hepaticoenterostomy or hepaticoduodenostomy; both in substitution to earlier practices of cyst enterostomy. Unexcised cysts are associated with higher incidences of carcinoma in the biliary tract and, also, with potential postoperative complications, such as cholangitis and lithiasis.[7-9]

Objectives

To assess results of the surgical treatment of choledochal cyst at our hospital, and to report symptoms observed and additional exams used in the diagnosis.

Patients and methods

We carried out a retrospective analysis of medical records of 18 patients submitted to surgical treatment of choledochal cyst between 1987 and 1997, related to the discipline on Pediatric Surgery at the Medical Sciences School of the Universidade Estadual de Campinas. The age of patients at diagnosis varied from 20 days to 13 years (average of 3 years and 8 months).

Choledochal cysts were classified according to Todani et al. (Table 1). Surgical treatment consisted of cyst excision with Roux-en-Y hepaticojejunostomy, with the exception of 1 patient with choledochal cyst type V. The patient with choledochal cyst type V had been treated with Roux-en-Y hepaticojejunostomy after the resection of a stenotic segment in the common hepatic duct. One patient had been submitted to cystoduodenostomy elsewhere and remained symptomatic. All patients had been submitted to cholecystectomy. Concomitant hepatic biopsies were carried out in 14 patients. After being released from the hospital, outpatient follow-up was carried out periodically.

Results

Out of the 18 patients submitted to surgical treatment of choledochal cyst, 3 were males and 15 were females, with a male-to-female ratio of 1:5. Table 2 presents distribution of patient age at diagnosis, in which 15 (83.4%) patients presented younger than 7 years of age.

Time between the onset of symptoms and diagnosis varied from 4 days to 7 years, with an average of 13.2 months. Prenatal diagnosis was not performed in any of the 18 patients. One patient had been submitted to cystoduodenostomy elsewhere, and was experiencing recurrence of symptoms. Jaundice was the most common initial symptom, present in 15 (83.3%) patients. Other symptoms observed were: dark urine (11 patients; 61.1%); acholic feces (10 patients; 55.6%); abdominal pain (9 patients; 50%); vomiting (8 patients; 44.4%); hepatosplenomegaly (4 patients; 22.2%); and abdominal mass (2 patients; 11.1%). Two patients presented with cholangitis, two with bile duct stones, and one with acute pancreatitis. Associated pathologies were congenital cardiopathy, intestinal malrotation, *pancreas divisum*, and cryptorchidism, each diagnosed in one patient.

Diagnosis was confirmed by sonography of the abdomen in 14 of the 18 patients (77%). CAT scan of the abdomen was carried out in nine patients in order to corroborate diagnosis. Endoscopic retrograde cholangio-pancreatography was carried out in four patients, indicating choledochal cyst and abnormal common hepatic duct (greater than 15 mm) in three of them. Endoscopic retrograde cholangiopancreatography was indicated for the following

Table 1 - Classification of cysts according to Todani et al.

Type of cyst	Classification
I	Common type: (a) choledochal cyst in a narrow sense; (b) segmental choledochal dilatation; and (c) diffuse or cylindrical dilatation
II	Diverticulum type in the whole extrahepatic duct
III	Choledochocele
IV	Multiple cysts at the intra and extrahepatic ducts
V	Single or multiple intrahepatic bile duct cyst (Caroli's disease)

Table 2 - Distribution according to age at diagnosis

Age	Number of patients	%
<1 year	5	27.8
1-3 years	5	27.8
3-5 years	3	16.7
5-7 years	2	11.1
>7 years	3	16.7
Total	18	100

patients: one 4-year old patient with type II choledochal cyst and idiopathic acute pancreatitis; two patients with sonography indicating gallstones and dilatation of the biliary tract; one patient with previous cystoduodenostomy carried out elsewhere.

Table 3 presents the distribution of choledochal cyst types, with none type-I patients, one type-II patient, seven type-IV patients, and one type-V patient, according to Todani's classification.[7] One case of choledochal cyst type I was associated with atresia of the terminal duct of the choledochus, diagnosed in the prenatal period. The patient with choledochal cyst type V presented stenosis of approximately 5 mm in the common hepatic duct, with inward cystic dilatation of the intrahepatic biliary system. These last two patients presented lethal complications during the postoperative period. The first patient died of disseminated intravascular coagulation during the early postoperative period. The second patient died of cholangitis and hepatic failure after the third postoperative appointment. Mortality following surgery was thus of 11%. Liver biopsies of both patients presented anatomicopathological diagnosis of biliary cirrhosis.

The hepatic implication observed in patients submitted to liver biopsy ranged from minimal portal reaction to varied levels of portal fibrosis and biliary cirrhosis. Follow-up was carried out with one patient after the first postoperative appointment. Fifteen patients were submitted to follow-up for periods ranging from 5 months to 9 years and 8 months (average of 3 years), out of which 13 (86%) were normal at hepatic function testing, at enzyme testing, and presented no history of cholangitis. Two patients presented chronic pancreatitis; one of these two patients also presented cholangitis and cryptorchidism; the other presented *pancreas divisum*. The latter patient was initially submitted to cystoduodenostomy elsewhere, and presented with recurrence of symptoms (abdominal pain and cholangitis). This patient was reoperated using cyst excision with Roux-en-Y hepaticojejunostomy at our hospital (Table 4).

Table 3 - Frequency of choledochal cyst types

Type of cyst*	Number of patients	%
I	09	50
II	01	05
IV	07	38
V	01	05
Total	18	100

*According to Todani et al.

Discussion

In 1959, Alonzo-Lej presented two cases of choledochal cyst and analyzed 94 patients described in the literature until that date. The author suggested the first classification for choledochal cyst including only extrahepatic dilatation.[6] Subsequently to the development and improvement of diagnostic methods, other authors added two new types of classification for patients with intrahepatic cystic dilatation.[7]

Choledochal cyst is considered a rare pathology that occurs more frequently in female pediatric patients. Also, choledochal cyst presents a higher incidence among the Eastern-hemisphere population. The estimated incidence of choledochal cyst in the Western-hemisphere population is of 1:100,000 to 150,000 live births.[1] In our study, 83.4% of the patients with choledochal cyst were younger than 7 years of age, and there was a prevalence of female patients (male-to-female patients ratio = 1:5).

There are many hypotheses as to the origin of choledochal cyst. The most accepted theory is that choledochal cyst may be related to an abnormal junction between the pancreatic duct and the common bile duct, allowing reflux of pancreatic juice into the bile duct. Presence of pancreatic juice and enzymatic activity on the epithelium of the biliary tree could be responsible for inflammation of biliary tract, which would result in degeneration of the bile duct wall. An abnormal junction can be found in up to 80% of the patients with choledochal cyst.[2-4] Reflux of pancreatic juice into the biliary tree has been demonstrated by levels of biliary amylase measured at operation[10] and by intraoperative biliary manometry.[4] Longstanding inflammation of the biliary tract caused by the reflux of pancreatic juice might be one of the factors in the carcinogenesis of the biliary tract.[4] Abnormal hepatopancreatic duct junction was demonstrated in three of four patients submitted to endoscopic retrograde cholangiopancreatography.

The classic triad (abdominal pain, jaundice, and abdominal mass) was not observed in our patients. The most frequent symptom was jaundice (83%), leading to functional obstruction (dark urine in 61%, and acholic feces in 55% of

Table 4 - Results of surgical treatment in 18 patients with choledochal cysts

Patient	Sex	Age	Cyst type	Procedure	Hepatic biopsy	Evolution	Duration of follow-up
01	F	01m 24d	IV	A*	No	Positive	09y 08m
02	M	01y 05m	I	A	Chronic cholangitis Mod. portal fibrosis	Positive	11m
03	F	07m	IV	A	Cholangitis Portal fibrosis	Chronic hepatitis (sonography)	03y 09m
04	M	01y 08m	IV	A	No	No follow-up	0
05	F	20d	I	A	Biliary cirrhosis	Death due to postoperative complication	0
06	F	02m	I	A	Biliary cirrhosis	Chronic hepatitis (sonography)	07y
07	M	09y 08m	V	Roux-en-Y hepaticoenterostomy	Biliary cirrhosis	Death due to postoperative complication	0
08	F	48d	I	A	Portal fibrosis	Cholangitis Chronic pancreatitis Cryptorchidism	06y
09	F	02y 08m	I	A reoperation	Minimal portal reaction	Chronic pancreatitis Cholecystoduodenal fistula	02y
10	F	03y 03m	IV	A	Minimal portal-biliary reaction	Positive	03y 06m
11	F	04y 05m	I	A	No	Positive	12m
12	F	13y	I	A	Mod. portal fibrosis	Positive	02y 04m
13	F	05y 10m	IV	A	Minimal portal-biliary reaction	Positive	01y 05m
14	F	06y 08m	IV	A	Intense portal reaction	Positive	02y
15	F	07y 07m	I	A	No	Positive	05m
16	F	02y 03m	IV	A	Portal fibrosis	Positive	01y 08m
17	F	02y 01m	I	A (removal of Laad's band)	Biliary cirrhosis	Positive	01y 05m
18	F	04y	II	A	No findings	Positive	

* A = Cyst excision with Roux-en-Y hepaticojejunostomy.

the patients). Abdominal pain and vomiting occurred in 50% and 44% of the patients, respectively. Abdominal mass occurred in just 11% of the patients. These results are similar to those found in the literature.[11,12]

Prenatal diagnosis of choledochal cyst can be carried out using sonography, and has been recently reported by other authors.[10,12] Postnatal confirmation of the diagnosis can be carried out using abdominal sonography, abdominal CAT scan, hepatobiliary scintigraphy with technetium-99 m, endoscopic retrograde cholangiopancreatography, and percutaneous transhepatic cholangiography.[13] In this sense, sonography should be carried out in children with suspicion of choledochal cyst, especially in cases of obstructive jaundice. Sonography diagnosed choledochal cyst in 77% of our patients. Abdominal CAT scan is considered the most accurate study for the examination of the intrahepatic biliary tree and the pancreas.[13] We have not been employing hepatobiliary scintigraphy or percutaneous transhepatic cholangiography, but we agree that they may be useful in specific cases. Endoscopic retrograde cholangio-pancreatography allows visualization of the biliary tree and pancreatic duct, as well as the identification of abnormalities. Endoscopic retrograde cholangio-pancreatography, however, may cause certain complications, such as cholangitis.[14] Though certain authors consider preoperative endoscopic retrograde cholangiopancreatography

indispensable, we only submitted certain patients to this examination because of its invasive characteristic. Four patients were submitted to endoscopic retrograde cholangiopancreatography: two patients who presented concomitant gallstones, one patient who presented acute pancreatitis (submitted to unsuccessful sonography and CAT scan), and one patient who had been submitted to cystoduodenostomy elsewhere. Intraoperative cholangiography was carried out in eight patients for the examination of anatomical conditions.

The most frequent choledochal cyst types among our patients were type I and IV, in 9 (50%) and 7 (38%) patients, respectively. There was only one patient with choledochal cyst type V (Caroli's disease), and one patient with choledochal cyst type II. These results were similar to those found in the literature.[11,12,14]

Hepatic lesions demonstrated by biopsy varied from minimal reaction to portal fibrosis. Patients with longer evolution of the disease presented higher incidence of hepatic lesions, probably resulting from extensive periods of biliary stasis and cholangitis.[12] Regression of hepatic fibrosis and normalization of the hepatic function have been shown in relation to early diagnosis of choledochal cyst.[15] In our study, we also observed varied levels of portal lesion, with four occurrences of biliary cirrhosis, probably due to the extensive period of time between the onset and the diagnosis of the disease (average of 13 months).

Different surgical treatments have been indicated for choledochal cyst. In the past, types I, II, and IV were treated with cyst enterostomy, which resulted in unsatisfactory rates of recurrent cholangitis and stenosis. In addition, the procedure of not removing the mucosa of the cyst with chronic inflammation could be related to the development of bile duct carcinoma.[19] Chijiiwa and Koga, in using cyst enterostomy, observed 80% of cholangitis and postoperative choledocholithiasis and hepatolithiasis. In that study, 70% of the patients needed reoperation.[11] Our study included a patient who had to be reoperated after having cysto-duodenostomy elsewhere, due to recurrent cholangitis and abdominal pain.

Currently, the treatment of choice consists of complete cyst excision with hepatoduodenal anastomosis, Roux-en-Y hepaticojejunostomy, or hepatic duct bifurcation.[4,11-16] Todani et al. did not find a significant difference in rates of stenosis and cholangitis between patients submitted to hepaticoduodenostomy or to Roux-en-Y hepaticojejunostomy.[17] Others have reported higher rates of complications related to Roux-en-Y, such as peptic ulcer, fibrosis, steatorrhea, and anastomotic dehiscence.[16-18] All our patients were submitted to Roux-en-Y hepaticojejunostomy, and none of the above complications were observed, except for steatorrhea, present in two patients with chronic pancreatitis. Two patients with previous hepatic cirrhosis died post-operatively due to complications related to deterioration of the hepatic function. Out of 15 patients who participated in long-term follow-up, 13 (86%) were doing well, did not present cholangitis, and presented normalization of the hepatic function.

In conclusion, data collected in this study indicate that the surgical treatment of choledochal cyst with cyst excision using Roux-en-Y hepaticojejunostomy presented satisfactory results in 86% of the patients. Poorer prognosis was related to patients who presented with serious deterioration of the hepatic function at diagnosis.

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