

## Outcomes of choledochal cysts with or without intrahepatic involvement in children after extrahepatic cyst excision and Roux-en-Y hepaticojejunostomy

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### ABSTRACT

**Background.** Type I and type IV-A choledochal cysts (CC) in Todani's classification are the most frequent types of CC. Unlike type I cyst, in which the dilatation is confined to the extrahepatic bile duct, type IV-A affects both extra and intrahepatic ducts. **Aim.** To review our experience of complete cyst excision with Roux-en-Y hepaticojejunostomy for the treatment of type I and type IV-A CC in childhood, in order to better characterize these entities. **Material and methods.** Data was collected retrospectively from a cohort of children who underwent cyst resection for CC from 1989 to 2011 in our institution. **Results.** Twelve patients were submitted to surgical excision of extrahepatic cyst and hepaticojejunostomy for treatment of type I (n = 6) and type IV-A (n = 6) cysts, with a complication rate of 25% (n = 3) and no mortality. Long term follow-up was available in 92% of patients, with a median of 10 years (2-22 years). Morbidities consisted of bile leak (2 patients) and late-onset cholestasis (1 patient); two of these required anastomotic revision. The results did not reveal any significant differences between the groups regarding postoperative outcomes (P > 0.05). Preoperative intrahepatic dilatation was found to permanently vanish in 83% of patients diagnosed with type IV-A cyst after operative repair. **Conclusions.** Intrahepatic dilatation of type IV-A cyst in children did not adversely affect the postoperative outcome after conventional surgical repair. This operative approach was effective in the management of type I and type IV-A cysts.

**Key words.** Choledochal cyst. Intrahepatic dilatation. Type IV-A choledochal cyst. Children. Cyst excision and Roux-en-Y hepaticojejunostomy.

### INTRODUCTION

Choledochal cysts are rare congenital entities characterized by single or multiple dilatations of the intra and/or extrahepatic biliary tree. Although the incidence in the western population is 1 in 100,000-150,000 live births, it is remarkably higher in Asian countries, particularly Japan, where they can be found in up to 1 in 1,000 live births. There is also an unexplained female:male preponderance, commonly reported as 1.5:1 in the pediatric population and in up to 4.9:1 in the adult population.<sup>1</sup> The etiology of choledochal cysts is still unclear, although

many theories have been proposed. Although they can present at any age, 75% of the diagnoses are made in childhood.<sup>2</sup>

Choledochal cysts are usually divided into several categories, based on anatomical findings. According to Todani's classification system,<sup>3-4</sup> choledochal cysts include five main types.

- **Type I.** Saccular or fusiform dilatation of a portion or entire common bile duct with normal intrahepatic duct.
- **Type II.** Isolated diverticulum protruding from the common bile duct.
- **Type III.** Dilatation of the intraduodenal segment of the common bile duct (choledochocele).
- **Type IV.** Multiple dilatations of the intrahepatic and extrahepatic biliary tract (IV-A) or multiple dilatations involving the extrahepatic biliary tree (IV-B), and
- **Type V.** Cystic dilatation of the intrahepatic biliary ducts (Caroli's disease).

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Manuscript received: March 16, 2011.  
Manuscript accepted: April 06, 2012.

Type I cyst is the most commonly found and represents 50-80% of all cases<sup>1</sup> and type IV was reported with incidences ranging from 5 to 83%, in different series.<sup>5-6</sup>

Clinical presentation of choledochal cysts varies and depends largely on the age of the child at presentation. The most common complaints of symptomatic patients are recurrent abdominal pain, obstructive jaundice or cholangitis. Abdominal mass is rarely found in children.

The potential morbidity and mortality of this condition is well documented in literature, namely the risk of biliary tract malignancies. This risk is diminished in children who present under the age of 10 years, where the overall risk is 0.7%.<sup>7</sup> A prompt and accurate diagnosis of choledochal cyst, followed by surgical or, rarely, endoscopical management, is therefore essential.

The traditional open procedure for the management of type I and type IV choledochal cysts is the complete excision of the extrahepatic cyst with biliary-enteric reconstruction through Roux-en-Y hepaticojejunostomy (HJ). Although this surgical approach does not completely eliminate morbidity risk, it has been shown to considerably improve prognosis.<sup>8</sup>

Long-term follow up is recommended for possible late complications after surgical treatment, particularly in type IV-A choledochal cyst, which seems to be more problematic regarding long-term complications.<sup>5,9</sup> However, it is unclear whether the remaining intrahepatic duct dilatation can really affect the postoperative outcome in children.

Scientific literature lacks information on the particularities of this type of cyst such as its real incidence, clinical presentation and postoperative outcomes, namely the persistence or not of the dilated intrahepatic biliary duct following conventional surgical procedure.

Herein, we report our experience in treating choledochal cysts using complete cyst excision and reconstruction through Roux-en-Y HJ over 22 years. This study aimed to compare the postoperative outcome of patients with choledochal cyst confined to the extrahepatic biliary tract (type I group) to those who had a choledochal cyst with extra and intrahepatic biliary tract dilatation (type IV-A group). This study also aimed to investigate whether a preoperative dilatation of large intrahepatic ducts (left/right hepatic ducts) persists after extrahepatic cyst excision and Roux-en-Y HJ, and if this persistence influences the postoperative long-term outcome.

## MATERIAL AND METHODS

This retrospective study was conducted in strict accordance to confidentiality recommendations, following the review and approval of our Faculty of Medicine Review Board.

The study population consisted of all patients who underwent resection of an extrahepatic choledochal cyst and Roux-en-Y HJ from October 1989 to October 2011 at the Pediatric Hospital of *Centro Hospitalar e Universitário de Coimbra* in Portugal. We included in this study all cases where choledochal cyst was the definite diagnosis, the surgical approach was a complete extrahepatic cyst excision with Roux-en-Y HJ and the surgery was performed at our hospital. All choledochal cysts associated to biliary atresia, treated by a different approach or operated at a different hospital were excluded.

Data was collected through retrospective review of the patient medical records. Patient demographics, presenting symptoms, preoperative complications of the disease, imaging studies, size and anatomical type of the cyst, plasma liver biochemistry, surgical repair, operative and postoperative morbidity, mortality, length of hospital stay, pathology and follow-up data were collected. All available pre or perioperative imaging information, which consisted of preoperative abdominal ultrasound, magnetic resonance cholangiography, abdominal computed tomography and intraoperative cholangiogram, was prospectively reviewed separately by two radiologists (H.M. and S.B.) and the patients were divided into two groups; type I group included patients who had extrahepatic choledochal cysts with normal intrahepatic duct and type IV-A group included patients who had both intrahepatic and extrahepatic bile duct dilatation. Disease outcome measures included early and late postoperative morbidity, and overall mortality. A secondary outcome measure was the postoperative fate of preoperative intrahepatic duct involvement. These data were gathered from the follow up abdominal ultrasounds.

Intrahepatic duct involvement was defined either as visualization of intrahepatic ducts on abdominal scan or visualization of a moderate to important intrahepatic duct dilatation on cholangiography imaging. Scans were regarded as "normal" if the ducts were not visualized.

For descriptive analysis, median with range were calculated from the numeric variables and numbers (percentage) were used to summarize demographic and practice data. Continuous variables were compared using the Mann-Whitney test and categorical

data using the Fisher's exact test.  $P < 0.05$  was considered significant. Statistical analysis was performed using SPSS version 17.0 (SPSS, Chicago, IL, USA).

## RESULTS

### Patient demographics and clinical characteristics

Twelve patients underwent total extrahepatic cyst excision and Roux-en-Y HJ at our hospital for choledochal cyst confined to the extrahepatic biliary tract (type I group) or association of intrahepatic and extrahepatic biliary tract involvement (type IV-A group), between October 1989 and October 2011. Table 1 summarizes the patient demographics and clinical characteristics of the two groups.

All children but one (of African descent) were Caucasian, 11 females (92%) and 1 male with a median age at surgery of 4.2 years (range 1.1-9.3 years). The median age at the onset of symptoms was 2.4 years and the median age at clinical suspicion of choledochal malformation was 2.5 years.

In one child the choledochal cyst was an incidental finding on an abdominal scan performed for an unrelated condition. The remaining 11 patients were symptomatic prior to diagnosis; presenting symptoms included jaundice in 5 (45%), recurrent abdominal pain in 3 (27%), abdominal mass in 1 (9%), and serious complications in 2 (18%); of these latter, one child presented with cholangitis and the other with peritonitis caused by cyst rupture.

Three of the previously jaundiced patients experienced cholangitis or pancreatitis while waiting for definitive repair; another patient with abdominal mass presented with cholangitis after cystoduodenostomy performed as a primary surgical procedure.

Overall, 6 (50%) episodes of serious complications occurred preoperatively.

All patients had available imaging information before cyst resection, which consisted of more than one imaging study. The 12 patients underwent preoperative abdominal ultrasound, which was complemented by magnetic resonance cholangiography in 6 (50%), abdominal computed tomography in 1 (8%) intravenous cholangiography in 1 (8%) or intraoperative cholangiogram in 8 (67%). Intrahepatic duct dilatation was confined to the first-order hepatic ducts in 5 of 6 patients. The remaining patient showed dilatation of first-order hepatic ducts and also moderate fusiform bilobar dilatation of the peripheral ducts.

Three (25%) of the patients underwent previous abdominal surgeries at our hospital consisting of neonatal repair of duodenal atresia in the first, open appendectomy and biliary external drainage (two separate laparotomies) in the context of choledochal cyst perforation in the second and cystoduodenostomy as primary surgical procedure in the third.

The surgical repair was performed by two pediatric surgeons (MFL and AR) and consisted of elective complete cyst excision with Roux-en Y HJ. The extrahepatic cystic bile duct, including gallbladder, was totally excised through laparotomy. The typical reconstruction involved a 30- to 40- cm Roux loop with a wide hepaticojejunostomy at the level of the common hepatic duct. Choledochoscopy was not part of our operative protocol. A liver biopsy was routinely performed.

According to the modified Todani classification, association of intrahepatic and extrahepatic biliary tract involvement (type IV-A cyst) was found in 6 (50%) of all cases before operative repair; the remaining 6 (50%) presented with choledochal cysts confined to the extrahepatic biliary tract (type I cyst).

**Table 1** Demographic and clinical characteristics of 12 patients with cyst excision and Roux-en-Y Hepaticojejunostomy over 22 years.

Characteristics	Type I cyst (n = 6)	Type IV-A cyst (n = 6)	Total (n = 12)
Female gender, n (%)	5 (83)	6 (100)	11 (92)
Median (range) age at surgery (years)	5.2 (1.8-7.6)	3.7 (1.1-9.3)	4.2 (1.1-9.3)
Median (range) age at initial symptoms (years)	3.1 (0.1-6.9)	2.2 (1-6.8)	2.4 (0.1-7)
Median (range) age at diagnosis (years)	3.2 (0.1-7.5)	3.4 (1-9.3)	2.5 (0.1-9.3)
Previous operation, n (%)	1 (20)	2 (33.3)	3 (25)
Pre-operative serious complications, n (%)	1 (20)	5 (83.3)	6 (50)
Median (range) choledochal cyst diameter (cm)	2.4 (0.8-3.4)	2.1 (0.7-7)	2.2 (0.7-7)
Median (range) post-operative follow-up (years)	6 (0-10)*	13.5 (4-22)*	9 (0-22)

P statistical significance value. \*P = 0.024.

The median extrahepatic cyst diameter was 2.2 cm (range, 0.7-7 cm), with no significant differences between the groups.

The median postoperative follow-up time was 9 years (0 to 22 years). Eleven patients (92%) had long-term follow-up (median 10 years, 2 to 22 years) and had liver biochemistry tests and ultrasound scans performed at least annually, as part of the scheduled postoperative follow-up. The remaining patient had a short follow-up since his surgery was performed one month before the end of this study.

Histological assessment confirmed choledochal cyst in all cases with no evidence of malignancy. Liver pathology results of specimens collected perioperatively were available in 10 cases (83%): portal fibrosis was found in 3 and secondary biliary cirrhosis in 1 patient. A follow-up liver biopsy was not regularly performed for evaluation of these lesions.

The small numbers limit the value of statistical analysis but no obvious intergroup differences in demographics and preoperative clinical characteristics were seen, namely in gender, age at clinical presentation, diagnosis and surgery, preoperative complications, liver histological characteristics and extrahepatic cyst size. However, patients in the type IV-A group showed a trend towards worse clinical preoperative disease and showed significantly ( $P = 0.024$ ) longer overall postoperative follow-up time (median and range: 13.5 years, from 4 to 22 years) than type I group patients (median and range: 6 years, from 0 to 10 years); again, long-term follow-up time was significantly longer ( $P < 0.044$ ) in the type IV-A group (median and range: 13.5 years, from 2 to 22 years) compared to type I group (median and range: 7 years, from 2 to 10 years).

### Postoperative complications

The overall postoperative complication rate after cyst excision and Roux-en-y HJ was 25% (3 of 12) and there was no mortality.

The median hospital stay was 8.5 days (range: 5-26 days). Perioperative complications occurred in 2 patients (16.6%), which consisted of bile leak from the hepaticojejunostomy; one child had a bile leak which healed spontaneously, and another child had a bile leak that required surgical revision.

During a median postoperative follow-up of 10 years (range 2-22 years), a third patient (9.1%) with long-term follow-up evidenced a late-onset complication: seven years after a successful cyst excision and Roux-en-Y HJ this child was found to have intrahepatic bile stasis, biliary stones and cholangitis, which

was later successfully repaired by surgical revision with hilar ductoplasty. This patient remains symptom-free at 4.6 years after reoperation.

Postoperative morbidity did not differ significantly between groups. Type IV-A group showed similar proportions of both early and late complications (33.3 vs. 16.7% and 16.7 vs. 0%, respectively, as well as similar length of hospital stay (9.5 days vs. 7.5 days) compared with type I group. All these differences were not statistically significant.

### Intrahepatic duct dilatation pre/post surgery differences

Regarding the fate of the intrahepatic dilatation of a type IV-A cyst, this dilatation was no longer permanently found in abdominal scan in 4 (67%) out of the 6 patients, following operative repair. Of the 2 patients with intrahepatic involvement found on their postoperative abdominal scans, one persisted with asymptomatic dilatation during the long-term follow-up (eleven years) and the other evidenced a late-onset recurrence of the intrahepatic duct dilatation; this dilatation later subsided after a successful anastomotic revision. Neither of the excised type I choledochal cysts presented postoperatively with intrahepatic duct dilatation.

## DISCUSSION

The main findings in the present study were:

- A high proportion of the type IV-A cyst (type IV-A/type I cyst of 1:1).
- Higher incidence of preoperative serious complications in type IV-A when compared to type I.
- Spontaneous postoperative extinction of intrahepatic duct dilatation and
- Lack of impact of preoperative intrahepatic dilatation on worse postoperative outcomes.

These findings expand previous insights into the meaning of pre-surgical intrahepatic dilatation of choledochal cysts in children.

In the present study we reviewed our experience in treating choledochal cysts, but we focused specifically on type IV-A cyst in Todani's classification, in order to study any potential adverse influence of the intrahepatic dilatation both on the clinical characteristics of the disease and the postoperative outcome. Additionally, other related issues which have been a matter of debate were addressed, namely the frequency of this type of cyst, the etiology and the fate of the



unexcised dilated intrahepatic duct in children operated using the conventional surgical technique.

In our hospital, a tertiary referral center for the care of complex pediatric diseases in the central region of Portugal, we have treated 12 patients with choledochal cysts, either type I or type IV-A in Todani's classification, over the past 22 years. These choledochal cysts were all managed by extrahepatic cyst excision and Roux-en-y HJ. This series includes data of a patient included in a previously published study by Bernardes, *et al.* that reported our experience with 3 patients who underwent cystoduodenostomy for operative repair of choledochal cysts over the twelve preceding years.<sup>10</sup>

Our results confirm the previously reported female preponderance, and this was demonstrated for both type IV-A and type I cysts.

Choledochal cysts presenting in childhood have been considered congenital in nature. In our series choledochal cysts were all also probably congenital, however the congenital origin can be called into question in a case of a neonate who had transitory cholestatic jaundice after duodenal surgery for congenital duodenal diaphragm and, although asymptomatic over the following years, was diagnosed with extrahepatic cystic dilatation.

In our study, most patients were symptomatic. A serious cyst complication, namely cholangitis, pancreatitis or cyst perforation, occurred frequently in the preoperative course of the disease, either as a presenting clinical feature or as recurrent or single episodic complication, accounting for a preoperative complication rate of 50%. Only one of the 12 patients, whose diagnose was an incidental finding, was completely free of symptoms; a second patient presenting with transitory neonatal cholestatic jaundice after operative repair of duodenal diaphragm remained symptom free over seven years, until the age of definitive diagnosis of choledochal cyst and subsequent operative repair. A trend for worse clinical presenting symptoms was seen among the patients with intrahepatic involvement.

In this cohort study, which includes only the two types of choledochal cysts (type I and type IV in Todani's classification) that had clear indication for surgical management through extrahepatic cyst excision and Roux loop reconstruction, we identified 50% type I and 50% in type IV-A cysts; this 1:1 proportion is highly unusual, but is in accordance to results found in some pediatric series.<sup>11,12</sup> As noted in the majority of choledochal cyst series, the most commonly reported type of cyst is type I in Todani's classification, representing up to 80% of all cases of

choledochal cysts, with type IV as the second most frequent, with the exact ratio being hard to determine.<sup>1</sup> Previous studies have suggested a large variation in the proportion of this latter type of cyst, varying from 5 to 83%.<sup>5-6</sup> These discrepant rates may be attributable to subjective criteria and individual variability on interpreting anatomical information used for classification or to specific characteristics in the studied population.

Preoperative diagnosis and classification of choledochal cysts can be made by standard imaging studies, including abdominal ultrasound, magnetic resonance cholangiography or intra-operative cholangiography. Abdominal ultrasound is invaluable in the evaluation of these lesions. Lately, magnetic resonance cholangiography has been widely used to assist in the diagnosis. This latter study and/or the intraoperative cholangiogram play a definite role in the planning of the choledochal cysts surgical repair. Abdominal CT has only limited utility in the diagnosis and preoperative planning in children, since malignancy rarely occurs at pediatric age.

In our patients, the classification of the choledochal cysts as type I or IV in Todani's classification was based on anatomical information provided both by all available preoperative imaging information and by intraoperative findings, including intraoperative and surgical records. Most of our patients had a pre/perioperative complete workup imaging evaluation. However, one of our 12 patients did not undergo any of the usual cholangiographic studies. This patient was our first case of choledochal cysts excision with Roux loop HJ; the child had had an intravenous cholangiography after unsuccessful cystoduodenostomy, at the beginning of our 22-year review, when magnetic resonance imaging modality was not as readily available; moreover, intraoperative cholangiography was not performed neither at the primary definitive surgical procedure (cystoduodenostomy) nor at the second procedure (cyst excision and Roux-en-y HJ).

Although we have considered both the above indicated imaging studies and the surgical records and all imaging results were analyzed blindly and separately by two radiologists, we cannot ensure its complete validity, given the retrospective nature of the present study and the lack of accepted objective criteria. Other authors, who have found that the distinction between Todani's type I and type IV cysts is arbitrary and artificial since the intrahepatic ducts are never completely normal, have reported similar uncertainties.<sup>13-14</sup> In particular, Visser, *et al.*<sup>13</sup> suggest that the numerical system of classi-

fication should be abandoned and the term congenital choledochal cyst should be restricted to the anomaly consisting of extrahepatic dilatation associated to variable degrees of intrahepatic dilatation. The other conditions (types II, III and V in Todani's classification) should be reassigned as choledochal diverticulum, choledochoceles and Caroli's disease, respectively. However, according to other authors, type I and type IV cysts are 2 separate entities and therefore should not be managed as a same condition.<sup>15</sup>

Given the potential for serious complications, namely anastomotic stricture and malignization,<sup>8,16</sup> cystoduodenostomy as definitive surgical treatment of choledochal cysts is no longer recommended; two out of three patients from our previous reported study, who underwent cystoduodenostomy as definitive repair, were later reoperated by cyst excision and a Roux-en-Y HJ. The remaining patient was lost to follow-up 3 years after the surgical procedure.<sup>10</sup> Instead, early cyst excision is the recommended procedure in order to avoid serious complications, such as malignant transformation. This latter one has been reported to occur in long-standing unexcised choledochal cysts.<sup>16</sup> All of our patients were managed by total cyst excision and the current study showed no evidence of malignant disease, either preoperatively or during the postoperative long-term follow-up.

Regarding the post-surgical fate of the intrahepatic duct dilatation found in preoperative imaging studies, five of the six cases with type IV-A cysts in Todani's classification had postoperative "normal" ultrasonography, with complete disappearance of the intrahepatic biliary dilatation; the remaining patient had an asymptomatic persistent intrahepatic dilatation seen on ultrasonography exams over the eleven-year follow up, although with no evidence of cholestasis. One of the patients that showed disappearance of the intrahepatic dilatation soon after the operative repair, later developed left intrahepatic duct dilatation complicated by bile stasis and stone formation.

Postoperative regression of the intrahepatic duct dilatation in type IV-A choledochal cyst is still a matter of debate. The present study suggests that spontaneous extinction of the intrahepatic duct dilatation in the type IV-A choledochal cyst occurs early postoperatively. However previous studies have presented conflicting evidence on the postoperative fate of the intrahepatic duct dilatation. In accordance with our study both Hill, *et al.*<sup>17</sup> and Ohi, *et al.*<sup>6</sup> have reported that intrahepatic duct di-

latation in type IV-A choledochal cysts in children diminishes or disappears after surgery. In their recently published series Hill, *et al.*<sup>17</sup> performed a retrospective review of children with choledochal cysts labeled as type IV (on imaging and at surgery) and who underwent surgery (excision of extrahepatic bile duct dilatation and hepaticojejunostomy). At 1-year postsurgery intrahepatic ducts diameter had decreased to normal values and had remained stable for up to a 10-year follow-up. These authors suggest that intrahepatic dilatation is related to sustained increase of intrabiliary pressure rather than any intrinsic intrahepatic cystic malformation and that effective surgery invariably reduces measured intrahepatic dilatation toward normal values. This opinion is also supported by the study of Ohi, *et al.*<sup>6</sup> Contradicting results have been reported by Koshinaga, *et al.*, in a series of congenital choledochal cyst with intrahepatic dilatation, where 79% of the cases showed postoperative intrahepatic dilatation persistence, frequently accompanied by ductal stenosis.<sup>18</sup>

Long-term complications include stone formation, recurrent cholangitis and malignant transformation.<sup>8</sup> According to Dabbas *et al.*, stone formation occurs in up to 5% of cases during follow-up.<sup>19</sup>

In the 11 patients with long-term follow-up we had one (9%) late complication. This child had a type IV-A cyst, accounting for a 17% complication rate within this group compared to 0% complications in type I group. Although there was a trend for a higher rate of late postoperative complications in our type IV-A group compared to the type I group, this difference was not statistically significant.

The already mentioned late-onset complication which occurred in a child of our type IV-A group is worthy of note. The patient, a 9-year-old girl, had undergone cyst excision and Roux-en-Y HJ when she was 22 months old. She remained asymptomatic for the following 7 years, with normal liver biochemistry tests and ultrasound scans. Seven years after surgery she presented with recurrent abdominal pain and, over the next few months, both bile stasis and stones in a dilated left hepatic duct were shown in hepatobiliary iminodiacetic acid (HIDA) scan and in abdominal ultrasound, respectively. After a cholangitis episode the child underwent a surgical anastomotic revision, including hilar ductoplasty. As no anastomotic stricture was found, a possible explanation for the bile stasis was an undetected stricture near the confluence of the hepatic ducts, which could be solved by hilar ductoplasty.<sup>20</sup> This situation, together with a known risk of 15-20% of postop-

perative complications,<sup>12,21</sup> stresses the need for adequate prolonged surveillance.

Abnormal liver histopathology, which was found in 4 out of 10 of our patients, also did not significantly influence the postoperative results; the 3 children with portal fibrosis and the child with biliary cirrhosis remained completely asymptomatic in the long-term follow-up, indicating that the relieve of biliary obstruction can sustain the progression of liver disease.

There are some limitations to the current study. It reports the experience of a single pediatric tertiary center of a western country. Furthermore, the race, ethnic composition, and other characteristics of our population together with a small population may limit wide application to other populations. However, our findings highlight important characteristics that warrant consideration for prognosis of our children with choledochal cysts undergoing conventional operative repair.

## CONCLUSIONS

This article expands current literature on outcome of choledochal cysts with or without intrahepatic dilatation in children. It further supports the recommended approach of choledochal cyst excision and Roux-en-Y hepaticojejunostomy as a safe and effective surgical technique in the management of type I and type IV-A choledochal cysts.

As we highlighted earlier in the discussion, among major findings in the present study, firstly we report the apparent lack of impact of a preoperative intrahepatic duct dilatation either on preoperative or postoperative outcomes. Although there was a trend for a higher complication rate, both pre and postoperative, in IV-A cysts, this anatomical characteristic did not show any evidence of significant clinical risk, even over long-term follow-up. Secondly, our study findings documented early postoperative spontaneous extinction of the intrahepatic duct dilatation of the type IV-A choledochal cysts. Lastly, our data/results suggest that classification of types I and IV choledochal cysts according to Todani's classification system may present a diagnostic dilemma in children and supports other previous studies that point to the belief that his latter category may be a transitory pressure-related result and does not always represent intrahepatic bile duct disease.

## ABBREVIATIONS

- **HJ:** hepaticojejunostomy.

## ACKNOWLEDGEMENTS

We are grateful to Dr. Ana Brett (Pediatrician), a native English language speaker, for her assistance in reviewing this article.

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