



# A narrative review of the modern surgical management of pediatric choledochal cysts

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**Abstract:** Choledochal cysts (CC) are rare congenital dilations of the biliary tract that harbor lifelong malignancy risk. CC are treated with surgical excision and bilioenteric reconstruction. In the modern era, the surgical approach to pediatric patients has enjoyed significant innovation with regards to minimally invasive techniques. In this review, we discuss these advances, including laparoscopic, single-incision laparoscopic, and robotic strategies, with a focus on the clinical outcomes of patients undergoing these procedures. By presenting an overview of the technical pearls emphasized by pioneers of these procedures, we examine the benefits and limitations of various minimally invasive techniques and analyze the utility and effectiveness of laparoscopy and robotics in comparison to each other and open techniques. Additionally, we highlight the importance of surgeon experience and skill in the management of this rare pediatric disease and explore the significance of the surgical learning curve in minimally invasive approaches in the excision of CC. We discuss the challenge of achieving surgical competency along this learning curve, and present proposed strategies to improve skill sets in the face of low case volumes. Finally, the relative dearth of data discussing long-term follow-up in these patients is discussed, and additional research regarding outcomes, malignancy risk and surveillance, and quality of life is necessary to better understand this disease and the implications of its surgical management.

**Keywords:** Choledochal cyst (CC); laparoscopy; minimally invasive; pediatric; robotic

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## Introduction

### *Epidemiology, pathogenesis and classification*

Choledochal cysts (CC) are a group of congenital cystic malformations that lead to dilation of the biliary tract. The incidence in the United States is rare at approximately 1 in 100,000 to 150,000 children; it is higher in East Asian populations with reports of 1 in 13,000 children being affected (1). Almost 80% of CC are now discovered

prenatally or in infancy, and are surgically addressed by complete resection and reconstruction of the bile drainage into the alimentary system.

Although several theories have been suggested for the etiology of CC, the “long common channel theory” appears to have been most accepted (2). This theory suggests that there is an anomalous union of the pancreatic and biliary ducts that may lead to increased reflux of pancreatic enzymes resulting in enzymatic injury of ductal wall with

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Normal	Type I	Type II	Type II	Type IV	Type V
	Extrahepatic A. Cystic B. Saccular C. Fusiform	Diverticulum	Choledochoceles	Intra/extrahepatic A. Both intra/extra B. Multiple extrahepatic only	Intrahepatic
	80–90%	2%	4–5%	10–15%	Rare

**Figure 1** The Todani classification of choledochal cysts was published in 1977 and amended in 2003 to add subtypes. Type I and type IV have the highest incidence.

consequent ductal dilation.

The most common classification utilized is the Todani classification, which expanded on the Alonso-Lej scheme. It groups the types of CC based on the location of the cyst and involved bile duct anatomy (3). Type I CC is characterized by dilation of the common bile duct, and is treated with cyst resection and hepaticoenterostomy. Type II is a diverticulum and can be managed with diverticulectomy; type III, a choledochoceles, is managed with endoscopic sphincterotomy. Type IV and V have multiple intra- and extrahepatic dilations and may additionally require liver resection or transplant. Type IVb CC (multiple extrahepatic cysts) are appropriate for cyst resection and hepaticoenterostomy, as is performed for Type I CC. In this review, we address the surgical approach for extrahepatic CC disease in type I and IV disease (*Figure 1*).

### Management rationale

The medical management of CC is limited to preoperative antibiotic treatment in the event of cholangitis or supportive therapy in the setting of pancreatitis. In the rare instance of spontaneous perforation, percutaneous drainage can help stabilize the patient prior to operative resection (4).

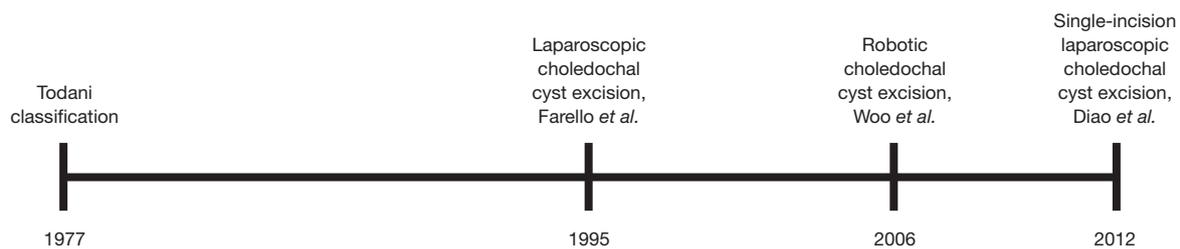
The potential of malignant degeneration to gallbladder carcinoma or cholangiocarcinoma is reported to be 6% to 30% in adults with CC, with a life-long malignancy risk of up to 4%, even after excision (5). The risk of

malignancy is higher in patients with anomalous pancreatic duct union, the anomaly thought to cause type I and IV CC (6). Therefore, the presence of a CC is indication for resection, and these children require long term follow-up for surveillance. While previous reviews have discussed the pathophysiology and general management of these lesions, there is no comprehensive narrative surrounding modern surgical management of CC. In this review, we focus on minimally invasive surgical techniques, surgical competency, and long-term follow-up in the approach to CC. Articles relevant to these topics were selected from a comprehensive search of PubMed database. Additional articles were identified from the references of selected literature.

We present the following article in accordance with the Narrative Review reporting checklist (available at <http://dx.doi.org/10.21037/tgh-20-235>).

### Surgical principles

Historically, cystenterostomy or a drainage procedure was the surgical approach for CC. This approach was found to be insufficient due to a high incidence of recurrence of symptoms as well as a relatively high incidence of malignant degeneration (7,8). As a result, cyst excision and bilioenteric reconstruction are the mainstays of surgical therapy. Two methods of reconstruction have been described: Roux-en-Y hepaticojejunostomy (HJ) and more recently, the hepaticoduodenostomy (HD).



**Figure 2** Timeline of minimally invasive surgical innovation in pediatric choledochal cyst excision. After the introduction of the Todani classification in 1977, it was not until 1995 when Farello *et al.* described the laparoscopic approach to pediatric choledochal cyst excision. Subsequently, the first case report describing the robotic strategy was published in 2006, followed by data examining single-incision laparoscopy in 2012.

## Minimally invasive approaches to CC in the pediatric population

### Laparoscopic approach

With the advent of laparoscopy, minimally invasive surgery (MIS) has strongly influenced the surgical management of CC (Figure 2). The first report to present the use of laparoscopic surgery to treat CC was published by Farello in 1995 (9). Since then, myriad articles promoting the feasibility, safety, and outcomes of this approach have followed. The safety of MIS versus open surgery was examined in a systematic review and meta-analysis published in 2015 by Zhen *et al.* (10). Seven observational, comparative studies were reviewed to compare outcomes between open and laparoscopic surgery for CC excision. This analysis encompassed 1,408 total patients, 611 of whom were managed with laparoscopy (43%). Meta-analysis was performed to detect differences in the following outcomes: operative time, hospital length of stay (LOS), length of time to return of bowel function, intraoperative transfusion, bile leak, abdominal bleeding, anastomotic stenosis, pancreatitis, adhesive small bowel obstruction and total complications. Laparoscopy resulted in significantly longer operative time, with a mean difference (MD) of 57 minutes (MD = 56.57; 95% CI, 32.20–80.93;  $P < 0.00001$ ). Conversely, LOS was approximately 2 days shorter in patients treated with laparoscopy (MD = -1.93; 95% CI, -2.51 to -1.36;  $P < 0.00001$ ), and time to recovery of bowel function was faster in patients who underwent laparoscopic surgery (MD = -0.94; 95% CI, -1.33 to -0.55;  $P < 0.00001$ ). Additional data supporting laparoscopy included lower rates of intraoperative blood transfusion (relative risk (RR) = 0.20; 95% CI, 0.11–0.38;  $P < 0.00001$ ) and adhesive intestinal obstruction (RR = 0.17, 95% CI, 0.04–0.77;  $P = 0.02$ ). Other

outcomes did not differ significantly between laparoscopic and open groups. Based upon these data, the authors concluded that laparoscopy is a safe and feasible approach to CC excision. Since the publication of this meta-analysis, additional data comparing laparoscopic and open approaches to pediatric CC management have emerged (11–17). Specific findings from these articles will be discussed below; generally, they support previous conclusions that laparoscopy is safe and effective to manage CC, and confers the benefit of decreased intraoperative bleeding and shorter hospital LOS despite an increase in operative time.

A large, international, multicenter study examined the implementation and outcomes of MIS approach to CC excision in adults and children over a span of forty years (11). Of their total cohort of 332 patients, 36 (11%) received an MIS approach (laparoscopic and robotic). Propensity matching revealed longer operative time (open 237 minutes versus MIS 301 minutes) and lower blood loss (open 50 mL versus MIS 18 mL) in the MIS group. There was no difference in cyst size upon pathologic examination (2.5 cm in open and MIS groups), but there were 3 cases of malignancy found in the open group (versus none in the MIS group). Postoperative LOS was 2 days shorter in MIS groups; complications occurred in 35% of the entire cohort within 30 days from surgery, with no differences in severity or frequency in complication in patients treated with open or MIS techniques. This study is unique in its presentation of CC management across the entire spectrum of patient age, and it supports the minimally invasive approach, especially for children. It is limited by the overall low implementation of MIS management of CC (approximately 11%), lacks long term follow-up data and may be biased by selection and retrospective nature (11). Longer follow-up and a large cohort are available from Qiao *et al.*, who

published their multicenter experience with 956 consecutive patients treated with laparoscopic CC excision, with a mean follow-up of 5.7 years (18). Complications were low overall, with 0.6% overall rate in anastomotic stricture, 1.5% bile leak, 0.8% Roux loop obstruction. There were two mortalities, including one in the immediate postoperative period due to bleeding. The operative time decreased as time from introduction of laparoscopy progressed, and centers who implemented the MIS approach later approached shorter operative times more quickly, perhaps from the collective experience of the study group.

Liem *et al.* reported the largest single-institution series of laparoscopic management of CC in pediatric patients to date (19). In their series, 400 patients were treated over a four year period with age ranges between one month and 16 years. Conversion to open and biliary leakage were rare. Compared to the HJ group, patients who underwent HD reconstruction had shorter operative times ( $220\pm 60$  vs.  $165\pm 51$  min), but similar LOS (approximately 6 days each). One of the major criticisms of the HD reconstruction has been a potentially higher risk of cholangitis. In this series, the rates of cholangitis were 1.5% and 0.6% for the HD and HJ groups, respectively ( $P=0.24$ ).

### *Laparoscopic excision in neonates*

Several studies have examined the safety of laparoscopic CC excision specifically in neonates. Liu *et al.* reported their experience with laparoscopic management of 9 neonates who presented with symptomatic CC (20). Four ports were used to perform the excision, and reconstruction was achieved with retrocolic Roux-en-Y hepaticojejunostomy in all cases, with the jejunojunction performed extracorporeally. The average age of the cohort was 24 days, with average weight of 3.7 kg and all presented with Todani type I lesions. There were no conversions to open and operative time was a mean of 216 minutes, which is comparable with other studies. Over a mean follow-up period of 32 months, there were no complications observed. The authors highlight the early consequences of untreated CC in symptomatic infants, given that biliary obstruction is generally high grade, and suggest that their data supports the feasibility and safety of early, MIS intervention (20).

With regards to the management of infants diagnosed with CC prenatally, Matsumoto *et al.* describe implementation of laparoscopy for excision and bilioenteric reconstruction (12). They present 7 patients in an open surgery cohort and 6 in a laparoscopic cohort. Laparoscopy

was performed in 4 asymptomatic infants when they reached 5 kg, and was performed earlier in 2 patients who were symptomatic from biliary obstruction (jaundice, acholic stools and hepatic stones). This small series again showed a significant difference in operative time between laparoscopic and open groups, with a mean of 92 additional minutes needed to complete laparoscopic excision. Blood loss was lower in the laparoscopic group, and they also had an earlier return to full feedings. There were no complications recorded after 33 months of follow up in the laparoscopic group.

Ryu *et al.* presented the largest series of neonatal patients, with 22 who received laparoscopic CC excision, compared to 21 who underwent open surgery (15). The surgical approach was similar to the method presented by Liu *et al.* (20). These authors show no significant difference in operative time, and similar outcomes with regards to hospital days and time to full feeding. Additionally, arterial carbon dioxide ( $\text{PaCO}_2$ ) level was measured intraoperatively and was not significantly higher in the laparoscopic patients, resolving concerns of hypercapnia in young patients receiving carbon dioxide insufflation. Parents were questioned regarding their satisfaction regarding the surgical wound and scar, and satisfaction scores were significantly higher in the parents of infants treated with laparoscopic CC excision (15).

Taken together, these articles suggest that laparoscopic CC excision is feasible and safe in neonates, whether symptomatic or asymptomatic and diagnosed on pre- or postnatal imaging. Limitations and benefits of MIS approach mirror those of older children, including longer surgical time, decreased blood loss, faster return of bowel function, and improved cosmesis.

### *Single-incision laparoscopy*

Further sophistication of the laparoscopic approach in CC management has been developed via single-incision techniques (21-24). The first description of single-incision laparoscopy for CC excision was published by Diao *et al.* in 2012. Nineteen patients underwent single-incision laparoscopic hepaticojejunostomy, two of whom were converted to conventional laparoscopy. Outcomes were compared to historical controls of patients who had received traditional laparoscopic hepaticojejunostomy for CC excision ( $n=218$ ). Operative time was similar, as were hospital LOS and time to full feeds (22). Building upon their initial data, a matched case-control study was then

performed to compare single-incision and conventional/four-port laparoscopy, with 75 patients included in each arm (these data overlap with and include patients from their initial report). Again, the authors found no difference in operative time, bleeding, hospital LOS, time to return of bowel function nor immediate or long-term outcomes. They report that single-incision patients had remarkable postoperative cosmesis with no visible scarring (25). A similar retrospective study from another institution examined single-incision outcomes in 17 patients who were matched to standard laparoscopy controls from a time period prior to implementation of single-incision laparoscopy. Again, this showed equivalent outcomes with possible improved cosmesis in single-incision cases (26). For all of the above studies, single-incision approaches were facilitated with transabdominal stay sutures for retraction, and jejunojunostomy was created extracorporeally with an end-to-side hepaticojejunostomy for biliary drainage. Overall, these data suggest that single-incision laparoscopy is an appropriate approach to pediatric CC excision, though the benefit of this technique is mainly cosmetic and appears to rely upon extensively on surgeon experience.

Diao *et al.* expound upon their single-incision experience specific to neonates in additional studies (focusing on both CC and cystic biliary atresia) (27), and this group's approach to giant CC (24) and perforated CC (21) are also available in the literature. Operative time and postoperative outcomes were acceptable and similar to their other reports, except in the group with perforation. Out of the cohort of 133 patients with perforated CC who underwent single-incision laparoscopic excision and hepaticojejunostomy, 15 (11%) were converted to open due to adhesions and bleeding. Three patients received blood transfusions and 2 had duodenal injuries which were repaired intraoperatively. The authors did employ a staged approach to children who presented with perforation and generalized peritonitis (n=52, 39%), with external biliary drainage performed prior to definitive cyst excision and bilioenteric reconstruction (21). This article highlights the difficult reality of perforated CC, since these children present with a much higher degree of intraabdominal inflammation and associated adhesions in the surgical zone, which confers higher operative risk. We surmise that the lowest-risk strategy to manage children with perforated CC is a patient-tailored approach with reliance upon preoperative drainage.

Other groups have subsequently published larger series describing the feasibility of single-incision laparoscopy for CC excision. In a group of 86 patients, this technique

was successfully used, with only 1% of cases requiring placement of additional ports (28). Overall, the data regarding single-incision laparoscopy for CC excision suggests that it is a feasible approach, with its main benefit being the improved cosmesis from trans-umbilical incision only. There are no randomized trials comparing single-incision with conventional laparoscopy, and existing studies are limited by small size, with questionable extrapolation of technique to other centers where existing experience may be lower. There is a lack of long-term follow-up to determine if cosmetic benefit remains present years after surgery. Additional, more rigorous comparisons with longer follow-up will better elucidate the usefulness of single-incision surgery for CC excision.

### *Robotic CC excision*

With regards to the application of robotic surgery to CC management, the earliest reports which described application of robotics to pediatric biliary surgery were published in 2006 (case report) and 2007 (small case series) (29,30). The case report from Woo *et al.* presented successful robotic excision of type I CC in a 5-year-old girl. The small case series from Meehan *et al.* presented 2 Kasai portoenterostomies and 2 CC excisions. Each of these studies were reconstructed with completely intracorporeal Roux-en-Y hepaticojejunostomy, and there were no reported complications. Subsequently, a larger case series of 14 children was published which highlighted the importance of surgeon skill versus the robotic learning curve (31). This group reported that the first 3 patients in their cohort experienced complications; these included conversion to open, anastomotic stricture requiring revisional surgery, and anastomotic leak treated with drainage. After the primary surgeon underwent robotic-specific skills training, the subsequent 11 cases were performed without incident (mean follow-up of 14.2 months), but operative time was long with a median of 570 minutes. This did decrease from 816 minutes in the first case to 432 minutes in the last case. Of note, the jejunojunostomy was performed extracorporeally and prior to robot docking while the hepaticojejunostomy was created with the robotic instruments.

Though initial reports regarding the potential for robotic intervention in the management of pediatric CC were unclear with regards to its role, a more robust, comparative study further elucidated the utility of robotics in this context. Kim *et al.* retrospectively compared open (n=43, 54%) and robotic (n=36, 46%) approaches. This group

also struggled with higher complication rates immediately after the introduction of robotic surgery (5 complications within 30 days in robotic group versus none in open group). These complications included anastomotic stricture, bile leak and intestinal obstruction. Robotic outcomes improved along with surgeon experience. It is notable that the robotic surgery patients were older (57 versus 30 months) and larger (19 versus 12 kg) (32).

More recent studies have shown improved outcomes compared to the early era of robotic CC excision. Pham *et al.* argue that the augmented dexterity conferred from robotic technology would lend itself to enhanced technique over laparoscopy (33). While this group did not compare their robotic experience to open or laparoscopic outcomes, their retrospective case series did present acceptable outcomes from robotic CC excision. Of the 39 patients they enrolled, none had any postoperative complications. Their operative time was significantly decreased compared to earlier data, at a mean of only 193 minutes. This group highlights the collaboration of a highly skilled robotic operative assistant and careful measurement of robotic arm interactions with adjustments as the study proceeded to optimize technique. Finally, a matched retrospective study comparing robotic versus laparoscopic anastomosis in pediatric CC excision was published by Koga *et al.* (34). These authors used a laparoscopic setup to perform initial excision and extracorporeal jejunojunostomy. Subsequently, 27 patients underwent laparoscopic hepaticojejunostomy while 10 received robotic hepaticojejunostomy. Robotic anastomoses were performed more quickly (70 versus 100 minutes) and surgeons perceived them to be easier to perform compared to laparoscopic anastomoses. However, the time benefit was lost with robotic setup and docking, since overall operative time was not significantly different between groups (618 minutes in robotic and 654 minutes in laparoscopic group). Overall, the operative time in this study was very high at an average of 10 to 11 hours per surgery.

Holistic examination of the data regarding robotic CC excision in children makes it clear that more research is needed to examine the utility of this approach. The most recent and larger studies do show acceptable complication rates, and robotic CC excision does not appear inferior with regards to complications if performed by experienced surgeons. On the other hand, concerns regarding the long operative times may be reinforced if forthcoming data shows similar trends; additionally, cost-effectiveness information should be reported given known expense of robotic platforms.

## Maintenance of surgical competency

CC are a classic example of a rare, anatomically complex congenital anomaly, which presents challenges to achieve sufficient maintenance of surgeon skill and experience. Surgical experience was perceived to be highly significant to successful innovation of new MIS approaches to CC excision, such as in single-incision laparoscopy. Additionally, data showed that both complications and operative time in robotic CC excision are significantly higher compared to laparoscopy at the onset, when overall experience with this technique is low. However, over time, surgeon experience increased with commensurate decrease in complication rates and operative times, which demonstrates a clear learning curve in the MIS management of pediatric CC (31). A learning curve is a measure of improvement in the technical performance of a certain procedure over time and is often defined in terms of the number of cases required to gain proficiency before a technique is considered optimally safe and effective to perform. In this section, we will examine the MIS learning curve in pediatric CC excision, discuss the data regarding challenges in skill maintenance of pediatric surgeons for this rare disease, and present proposed strategies to increase skill in light of low case volumes.

Though it is known that adverse events decrease with increasing experience, Wen *et al.* in 2017 set out to precisely define the number of cases to achieve the learning curve for laparoscopic CC excision (35). By prospectively reviewing 104 patients undergoing laparoscopic CC excision and Roux-en-Y hepaticojejunostomy performed by a single surgeon, they found that the learning curve for CC excision was approximately 37 cases. The cases were then evaluated by early experience (less than 37 cases) and late experience (the following 67 cases). Overall, they found a decrease in operative time (352 *vs.* 240 min;  $P < 0.001$ ), LOS (9.4 *vs.* 7.8 days;  $P = 0.01$ ) and complication rate (12.5% *vs.* 1.5%;  $P = 0.02$ ) with increasing numbers of procedures performed. A multivariate analysis was performed which found that completion of the learning curve (reaching 37 cases) and the amount of patient adhesions were independent predictors of operative time, perioperative adverse outcomes and LOS.

Wen *et al.* also evaluated the learning curve of the two major and most difficult steps of the procedure, cyst resection and creation of the hepaticojejunostomy, and found them to be 29 and 42 cases, respectively, concluding that the anastomosis time was key and could be shortened with practice. In addition to experience, the authors state that technical skills are reasons for failure of laparoscopic

CC excision and recommend training and simulation as well as supervision by more senior surgeons. Due to the association between outcomes and adhesions, the authors also suggest that surgeons who are early along in their learning curve implement careful patient selection, opting for patients with shorter disease duration.

In order to proceed along the learning curve, exposure and repetition are necessary. In direct opposition to this for diseases such as CC is the relative stable case volume and the increasing numbers of pediatric surgeons (36). The relationship between case volumes and outcomes has been widely studied and there is a positive correlation between higher case volumes and patient outcomes (37). As previously stated, CC is a rare condition in the United States, affecting approximately 1 in 100,000 to 150,000 children, making CC case volume and exposure difficult, particularly in areas of geographic isolation. In fact, from 2009 to 2013, 60% of board-certified pediatric surgeons reported no exposure (zero cases) to CC or biliary atresia (38).

While attending pediatric surgeons may find it difficult to maintain laparoscopic CC excision skills, trainees face an additional challenge. Trainees in both general surgery and pediatric surgery are required to perform “index cases,” and despite an 88% increase in pediatric surgery trainees, there has been no change in complex case volume (such as hepatic lobectomy, biliary atresia or CC excision) in pediatric surgery from 2005 to 2017 (36). Additionally, upon review of pediatric surgery fellow case records from 2000 to 2017, Park *et al.* found that there was at least 1 trainee per year who reported 0 complex CC or biliary atresia cases during their entire two years of pediatric surgery fellowship.

Sharing of cases may represent a helpful adjunct to improve exposure in lieu of increased clinical volume. A study by Johnson *et al.* in 2019 evaluated the implementation of a team-based approach to surgical care in order to increase exposure to rare index cases in a geographically isolated setting (37). A retrospective review of cases performed by at least 2 attending surgeons was conducted. The authors found that 100% of their CC excisions were team surgeries and, by utilizing a team-based approach, their case volume exceeded the national average for CC excision (1.5 *vs.* 0.9 per surgeon) (37). Of note, they also reported resident satisfaction with exposure to cases using a team-based approach. These findings suggest a team surgical approach may be a potential solution to increase experience per surgeon in support of maintenance of the skillset required for rare congenital diseases such as CC excision.

Simulation also plays a key role in acquiring and maintaining surgical skills. Surgical simulation focuses on the technical and cognitive aspects of a procedure and is a method for both trainees and surgeons to improve or maintain their surgical skills (38). Previous studies have found that simulation training leads to improvement in operative times, technical skills and patient-outcomes. Burdall *et al.* evaluated the use of a 3D-printed laparoscopic model for CC excision (39). Their model was tested by a group of ten pediatric surgeons, with interest in MIS, who rated the model 5.6/10 in tactile likeness, 6.3/10 in complexity and 7.4/10 in usefulness, suggesting a benefit of laparoscopic simulation with modeling. The authors also described additional benefits of 3D printing and simulation such as the ability to model variations in structure and allow for patient specific practice and operative planning.

Overall, simulation and team-based operations are feasible tactics to counter low case volumes and promote skills maintenance in pediatric surgeons who manage CC. What remains to be studied is whether or not these strategies may be linked to measurable improvements in surgeon skill or patient outcomes. Data is also needed to determine if robotic skills are transferrable from other pediatric or biliary conditions; aforementioned studies suggest that this may be true for CC and biliary atresia. Collectively, these data highlight a common challenge of the surgical management of complex, rare anomalies in children.

### Long-term outcomes and transition to adult care

Prognosis overall is excellent for CC managed with surgical resection. The risk of malignancy, even after complete operative resection, remains higher than the general population and can be as high as 4% (1,5). Groups have advocated for surveillance with serial CA 19-9 and abdominal ultrasound. However, more research is needed to devise the optimal follow-up and surveillance strategies is needed.

Post-operative complications not related to malignancy include anastomotic stricture, cholangitis, pancreatitis, bowel obstruction, and need for reoperation. In a retrospective review of 54 children who underwent laparoscopic CC excision and biliary-enteric reconstruction with HD versus Roux-en-Y hepaticojejunostomy anastomoses, it was found that there were no statistical differences in the rate of anastomotic leakage requiring reoperation, anastomotic stricture, cholangitis, abdominal

pain or gastritis, or need for reoperation (40). A Japanese single institution cohort of 110 patients with CC found that late complications were observed within 20 years and included intrahepatic bile duct dilation (0.9%), intrahepatic bile duct stones (2.7%), and adhesive bowel obstructions (3.6%). They did not have any malignancies in their series (41). While these data shine light on the long-term outcomes of patients who received CC excision, they are relatively small in size and scope. Higher volume, multi-institutional data is needed, to include quality of life information and follow-up well into adulthood, and would greatly benefit the understanding of this disease and its management.

## Conclusions

This review focuses on the modern surgical management of pediatric CC, specifically types I and IV. It has been well-established that all children with a diagnosis of CC require excision and hepatic drainage given the malignancy risk associated with this disease process. However, significant innovation has emerged with regard to the surgical approach of CC excision, specifically in MIS techniques. It is clear that the open surgical management of pediatric CC is no longer the standard of care; robust data confirms the safety of MIS to manage these anomalies. This is not surprising, as the field of pediatric surgery has always pushed the envelope of less invasive approaches in order to minimize the surgical trauma that our small, delicate patients must endure. Overall, the selection of MIS (laparoscopic, single incision versus robotic) or even of open techniques should be tailored to the individual patient. Amongst these approaches, a dearth of data supporting implementation of robotic technology in this disease process calls for caution in employing this specific modality, and larger, comparative studies should be carried out to further explore the utility of robotic interventions in pediatric CC. Pediatric surgeons with higher accumulations of experience with MIS management of CC should lead the care of these patients in order to achieve less invasive approaches while still ensuring optimal outcomes, with a defined need to study surgical competency and skills maintenance in the operative management of this rare disease. Finally, multi-institutional studies with follow-up into adulthood are required in order to fully understand the spectrum of this disease and its implications for long-term wellbeing after successful CC excision.

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