

Choledochal Cysts: Contemporary Diagnosis, Surgical Management, and Long-Term Surveillance

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ABSTRACT

Choledochal cysts, increasingly referred to as congenital biliary dilatation, are uncommon abnormalities of the biliary tree associated with biliary stasis, pancreatobiliary reflux, recurrent inflammation, pancreatitis, cholangitis, stone formation, progressive hepatobiliary injury, and an increased lifetime risk of biliary tract malignancy. Although many cases are diagnosed during childhood, adult presentation remains clinically relevant because delayed diagnosis is often associated with complicated disease, chronic inflammation, biliary stones, adhesions, and greater concern for cholangiocarcinoma or gallbladder carcinoma. Contemporary evaluation relies on ultrasonography, magnetic resonance cholangiopancreatography, computed tomography in selected cases, and therapeutic or problem-solving use of endoscopic retrograde cholangiopancreatography. Complete cyst excision with cholecystectomy and biliary reconstruction remains the cornerstone of treatment for most extrahepatic choledochal cysts. However, important controversies persist regarding the timing of surgery in prenatally diagnosed neonates, hepaticoduodenostomy versus Roux-en-Y hepaticojejunostomy, the role of laparoscopic and robotic surgery, and the optimal strategy for long-term surveillance. This narrative review provides a contemporary overview, updated through May 2026, of classification, pathogenesis, diagnostic evaluation, operative principles, reconstruction strategies, minimally invasive approaches, postoperative complications, malignancy risk, and follow-up.

KEYWORDS: choledochal cyst, congenital biliary dilatation, hepaticojejunostomy, hepaticoduodenostomy, biliary reconstruction, long-term surveillance

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INTRODUCTION

Choledochal cysts are congenital or developmental dilatations of the biliary tree that may involve the extrahepatic bile duct, intrahepatic ducts, or both. Although the term “choledochal cyst” remains widely used in clinical practice, contemporary hepatobiliary and pediatric surgical literature increasingly uses the term “congenital biliary dilatation” because these lesions are not always true cysts and may present as fusiform, segmental, diverticular, intraduodenal, intrahepatic, or combined ductal dilatations [1-4]. Their clinical relevance extends beyond anatomic dilatation, as they are associated with recurrent biliary inflammation, pancreatitis, cholangitis, biliary sludge or stones, progressive liver injury, postoperative complications, and malignant transformation [1,3,5,6].

The management of choledochal cysts has evolved considerably. Historical internal drainage procedures have largely been abandoned because they leave abnormal biliary epithelium in place and do not eliminate the long-term risks of recurrent cholangitis, stone formation, and malignancy [1,6-8]. Modern treatment is based on complete excision of the abnormal extrahepatic biliary segment, cholecystectomy, and biliary-enteric reconstruction, most commonly through Roux-en-Y hepaticojejunostomy [1,7,8]. However, hepaticoduodenostomy has re-emerged as an alternative in selected patients, prompting renewed discussion regarding reflux, endoscopic accessibility, anastomotic complications, and long-term outcomes [9,10].

Advances in minimally invasive surgery have also changed the operative landscape. Laparoscopic excision has demonstrated favorable outcomes in experienced centers, and

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robotic approaches have expanded the feasibility of precise biliary reconstruction in selected pediatric, neonatal, low-weight, and adult patients [9,11,12]. Despite these advances, the procedure remains technically demanding because it requires safe dissection within the hepatoduodenal ligament, careful distal duct management near the pancreas, complete cyst excision, and a durable biliary-enteric anastomosis [7,8,12,13]. Long-term surveillance remains essential because postoperative anastomotic stricture, cholangitis, intrahepatic stones, pancreatitis, and residual malignancy risk may occur even after technically successful surgery [12,14-19].

This review provides a contemporary synthesis, updated through May 2026, of choledochal cysts, with emphasis on contemporary diagnosis, surgical management, reconstruction strategies, minimally invasive and robotic approaches, postoperative complications, malignancy risk, and long-term surveillance.

METHODS AND LITERATURE SEARCH STRATEGY

A narrative literature review was performed using PubMed, PubMed Central, and Google Scholar. The search focused primarily on articles published between January 2020 and May 2026, while older landmark studies were considered when necessary for classification, terminology, or historical context. Search terms included “choledochal cyst,” “congenital biliary dilatation,” “Todani classification,” “pancreatobiliary maljunction,” “hepaticojejunostomy,” “hepaticoduodenostomy,” “laparoscopic surgery,” “robotic surgery,” “postoperative complications,” and “malignancy.” Priority was given to comprehensive reviews, systematic reviews, meta-analyses, pediatric and adult surgical series, technical studies, and long-term outcome studies. Additional open-access figures were selected to illustrate endoscopic, radiologic, surgical specimen, and biliary reconstruction findings relevant to this review [10,20].

Because this was designed as a narrative review rather than a systematic review, no formal protocol registration, risk-of-bias assessment, or quantitative synthesis was performed. Articles were selected according to clinical relevance, recency, methodological quality, and applicability to surgical decision-making. The manuscript was structured as a practical surgical update for general surgeons, hepatobiliary surgeons, pediatric surgeons, gastroenterologists, and clinicians involved in the multidisciplinary care of patients with choledochal cysts.

DEFINITION AND TERMINOLOGY

Choledochal cysts are congenital dilations of the biliary tree involving the extrahepatic bile duct, intrahepatic bile ducts, or both. The term remains clinically familiar, but “congenital biliary dilatation” more accurately describes the spectrum of disease because the abnormality may not always be cystic in morphology [1-4]. Some lesions are fusiform, others are saccular or diverticular, and others involve intraduodenal or

intrahepatic segments. This distinction has practical implications because disease morphology influences imaging interpretation, operative planning, reconstruction strategy, and long-term follow-up.

Terminology also varies geographically and by specialty. In pediatric surgery, “choledochal cyst” remains common, whereas Japanese and East Asian literature often uses “congenital biliary dilatation,” particularly when discussing pancreatobiliary maljunction, postoperative outcomes, long-term complications, and carcinogenesis [14-16,21-23]. For clinical purposes, both terms refer to a disease spectrum that requires accurate anatomic definition and, in most extrahepatic forms, definitive surgical treatment.

EPIDEMIOLOGY AND CLINICAL RELEVANCE

Choledochal cysts are uncommon, but their impact is significant because untreated disease may lead to recurrent biliary obstruction, cholangitis, pancreatitis, liver fibrosis, intrahepatic stones, and malignancy [1,3,5,6]. They are reported more frequently in Asian populations and show a female predominance, although cases occur worldwide and across all age groups [1,3,4,24]. Pediatric diagnosis is common, especially when symptoms occur early or when prenatal ultrasonography detects biliary dilatation [7,25,26]. However, adult presentation remains clinically important because delayed diagnosis is often associated with chronic inflammation, biliary stones, pancreatitis, adhesions, and malignancy risk [5,24,27,28].

Adult cases may differ substantially from pediatric cases. Children may present with jaundice, abdominal pain, vomiting, abdominal mass, cholangitis, or pancreatitis, whereas adults more often present with recurrent right upper quadrant pain, cholangitis, pancreatitis, choledocholithiasis, hepatolithiasis, abnormal liver tests, or incidental imaging findings [1,5,24,26]. In adults, diagnostic evaluation must also consider cholangiocarcinoma and gallbladder carcinoma, particularly in patients with longstanding symptoms, complex biliary anatomy, or imaging features suspicious for malignancy [5,21].

CLASSIFICATION

The Todani classification remains the most widely used anatomic system. Type I lesions involve cystic or fusiform dilatation of the extrahepatic bile duct and are the most common. Type II lesions are diverticula arising from the extrahepatic bile duct. Type III lesions, or choledochoceles, involve intraduodenal dilatation of the distal common bile duct. Type IV-A lesions involve both intrahepatic and extrahepatic ductal dilatation, whereas type IV-B lesions involve multiple extrahepatic ductal dilations. Type V disease, also known as Caroli disease, involves multiple intrahepatic bile duct dilations [1-4].

The classification is clinically useful because it guides diagnostic interpretation and operative planning (Figure 1). Type I and many type IV lesions are generally treated with

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complete extrahepatic cyst excision and biliary reconstruction [1,7,8]. Type III choledochoceles may require a different strategy, often involving endoscopic management in selected cases depending on anatomy, symptoms, and associated complications [2]. Type IV-A disease is more complex because intrahepatic ductal involvement may persist after extrahepatic cyst excision and predispose to cholangitis,

intrahepatic stones, hilar stenosis, and long-term complications [17,18,29]. Recent proposals for new classifications and laparoscopic treatment strategies highlight that traditional anatomic categories should be interpreted alongside ductal stenosis, intrahepatic disease, operative complexity, and long-term risk [30].

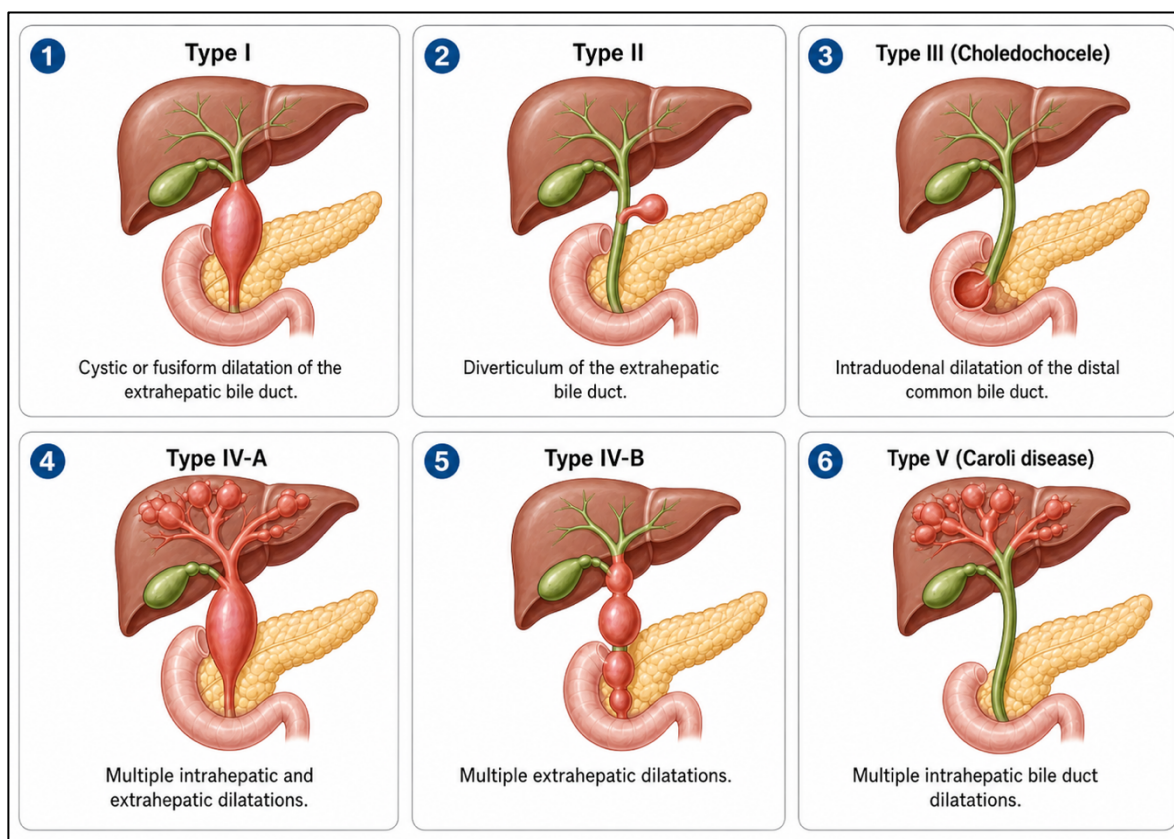


Figure 1. Todani classification of choledochal cysts. Schematic illustration of the Todani classification of choledochal cysts. Type I represents cystic or fusiform dilatation of the extrahepatic bile duct; type II represents a diverticulum of the extrahepatic bile duct; type III, or choledochocoele, corresponds to intraduodenal dilatation of the distal common bile duct; type IV-A involves both intrahepatic and extrahepatic ductal dilatation; type IV-B involves multiple extrahepatic ductal dilations; and type V, or Caroli disease, involves multiple intrahepatic bile duct dilations. This original schematic was created by the authors based on descriptions in the literature [1-4].

PATHOGENESIS AND MOLECULAR INSIGHTS

The pathogenesis of choledochal cysts is multifactorial. The most accepted mechanism involves an abnormal pancreatobiliary junction, which allows reflux of pancreatic enzymes into the biliary tree. This reflux may promote chronic epithelial injury, inflammation, weakening of the duct wall, and progressive biliary dilatation [1,3,7,31]. Biliary stasis and recurrent inflammation further contribute to cholangitis, stone formation, fibrosis, dysplasia, and carcinogenesis [1,6,21].

Recent molecular studies have expanded understanding of the biological mechanisms underlying congenital biliary dilatation. Studies of biliary epithelium suggest that carcinogenesis may involve genetic and epigenetic regulatory abnormalities, supporting the concept that chronic inflammation and molecular field changes contribute to

malignant transformation [21]. Genomic and transcriptomic analyses have identified potential pathways involved in choledochal cyst pathogenesis, while lncRNA-mRNA coexpression studies suggest that different subtypes may have distinct biological mechanisms [22,23]. Although these molecular findings are not yet routinely incorporated into surgical decision-making, they may eventually support risk stratification, postoperative surveillance, and individualized cancer-risk assessment.

Fibrosis is another clinically relevant component of the disease. Noninvasive models have been developed to predict advanced fibrosis in patients with choledochal cysts, reflecting the need to identify progressive liver injury before definitive surgery [27]. Fibrosis risk is particularly important in delayed diagnosis, type IV-A disease, recurrent cholangitis,

longstanding biliary obstruction, and prenatally diagnosed lesions that progress after birth [25-27,32].

CLINICAL PRESENTATION

Clinical presentation depends on age, cyst type, pancreatobiliary junction anatomy, associated inflammation, and presence of complications. The classic triad of abdominal pain, jaundice, and abdominal mass is historically described but is not present in most patients. Pediatric patients may present with jaundice, abdominal pain, vomiting, pancreatitis, cholangitis, failure to thrive, or abnormal liver tests [7,8,24]. Some cases are detected prenatally or during neonatal

evaluation, especially when ultrasonography identifies cystic dilatation in the porta hepatis [25,26]. Adults frequently present with less specific symptoms. Recurrent right upper quadrant pain, nausea, jaundice, pancreatitis, cholangitis, choledocholithiasis, hepatolithiasis, or incidental imaging findings may lead to diagnosis [1,5,24,26,28]. Because malignant transformation is more concerning in adults, any patient presenting with wall thickening, mural nodularity, mass-like enhancement, obstructive jaundice, unexplained weight loss, or suspicious laboratory findings requires careful oncologic evaluation [5,21].

Table 1. Pediatric versus adult presentation of choledochal cysts.

Feature	Pediatric patients	Adult patients
Usual timing of diagnosis	Prenatal period, neonatal period, infancy, or childhood	Delayed diagnosis, incidental discovery, or diagnosis after complications [1,5].
Common symptoms	Jaundice, abdominal pain, vomiting, cholangitis, pancreatitis	Right upper quadrant pain, cholangitis, pancreatitis, stones, abnormal liver tests [7,8].
Typical clinical concern	Progressive biliary obstruction, liver injury, pancreatitis, cholangitis	Complicated disease, adhesions, stones, malignancy risk [24-26].
Operative issues	Small duct diameter, timing of surgery, reconstruction quality	Chronic inflammation, difficult dissection, malignancy exclusion [26,28].

DIAGNOSTIC EVALUATION

Diagnosis requires integration of clinical presentation, laboratory findings, and imaging. Laboratory findings are nonspecific but may demonstrate cholestasis, hyperbilirubinemia, elevated aminotransferases, elevated alkaline phosphatase, elevated gamma-glutamyl transferase, or inflammatory markers during cholangitis. Serum amylase or lipase may be elevated in pancreatitis. In adult patients or those with suspicious imaging features, tumor markers such as carbohydrate antigen 19-9 may be considered, although interpretation should be cautious because inflammation and obstruction can also cause elevation [1,3,5]. Ultrasonography is commonly the initial imaging modality, especially in children and prenatally detected cases. It can demonstrate cystic or fusiform biliary dilatation, gallbladder abnormalities, intrahepatic ductal involvement, stones, and associated hepatic changes [7,25,26]. However, ultrasound is operator-dependent and may be limited in defining distal duct anatomy, pancreatobiliary junction anatomy, or complex intrahepatic involvement.

Magnetic resonance cholangiopancreatography (MRCP) is central to contemporary evaluation because it provides noninvasive delineation of the biliary tree, intrahepatic ductal anatomy, distal bile duct, pancreatic duct relationship, hilar stenosis, and associated pancreatobiliary anomalies [1,3,7,18]. MRCP is particularly important for operative planning in type IV-A disease, suspected hilar stenosis, intrahepatic stones, or complex ductal anatomy [18,29]. Computed tomography is useful in adults, complicated disease, suspected malignancy, pancreatitis, inflammatory changes, and preoperative staging when cross-sectional detail is required (Figure 3) [1,5,20,26]. Endoscopic retrograde cholangiopancreatography (ERCP) is no longer routinely required for diagnosis in most patients because MRCP provides excellent noninvasive anatomic characterization. However, ERCP remains useful in selected diagnostic or therapeutic circumstances, including choledochoceles, pancreatobiliary maljunction, biliary stones, cholangitis, postoperative strictures, and altered postoperative anatomy when combined with balloon-assisted techniques (Figure 2) [2,19,20,31].

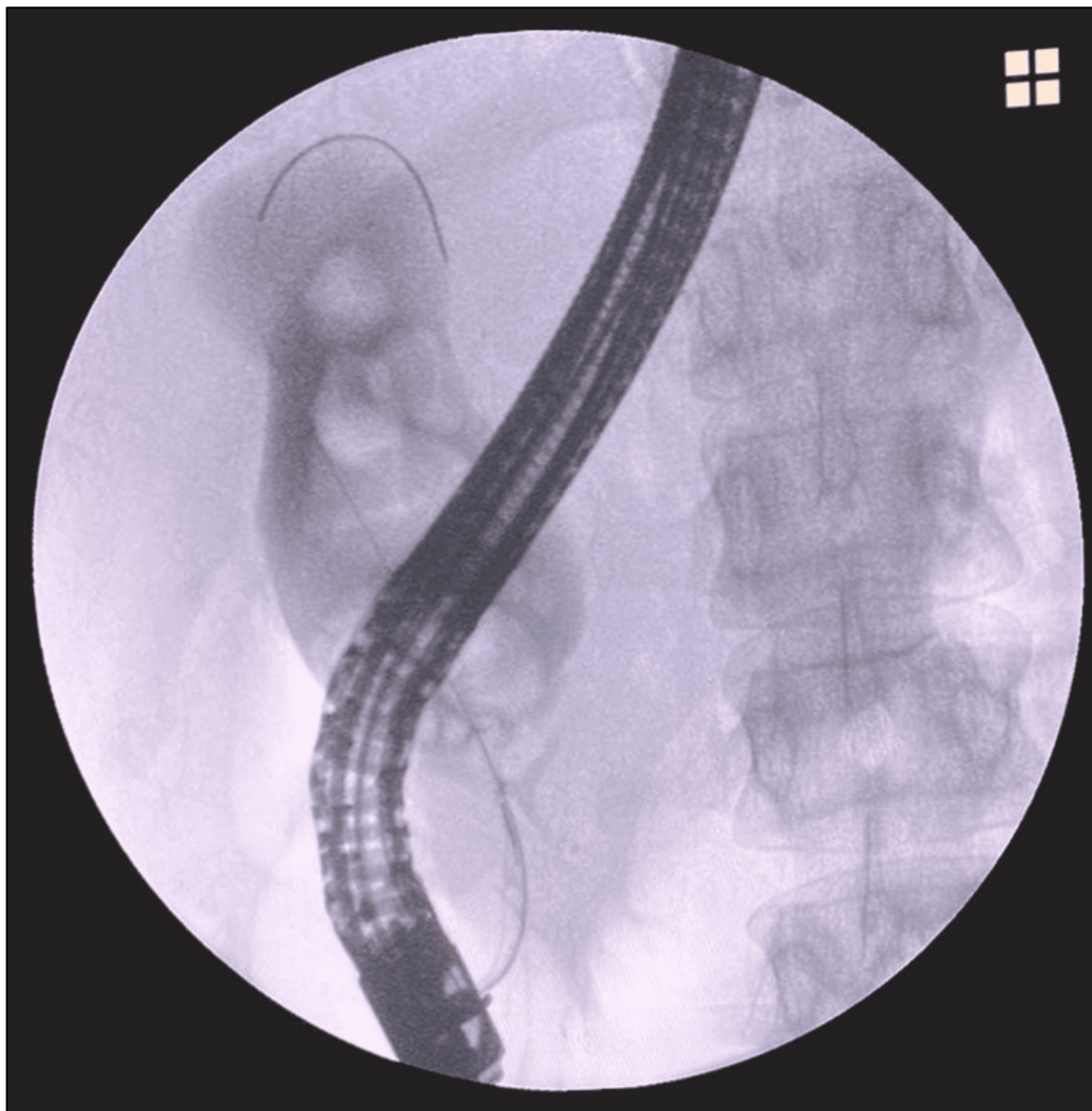


Figure 2. Endoscopic retrograde cholangiopancreatography demonstrating choledochal cyst with intraluminal filling defects. Endoscopic retrograde cholangiopancreatography showing contrast opacification of a markedly dilated common bile duct, with multiple intraluminal filling defects consistent with stones within the choledochal cyst. Reproduced from Villarino Zapata et al. [20].

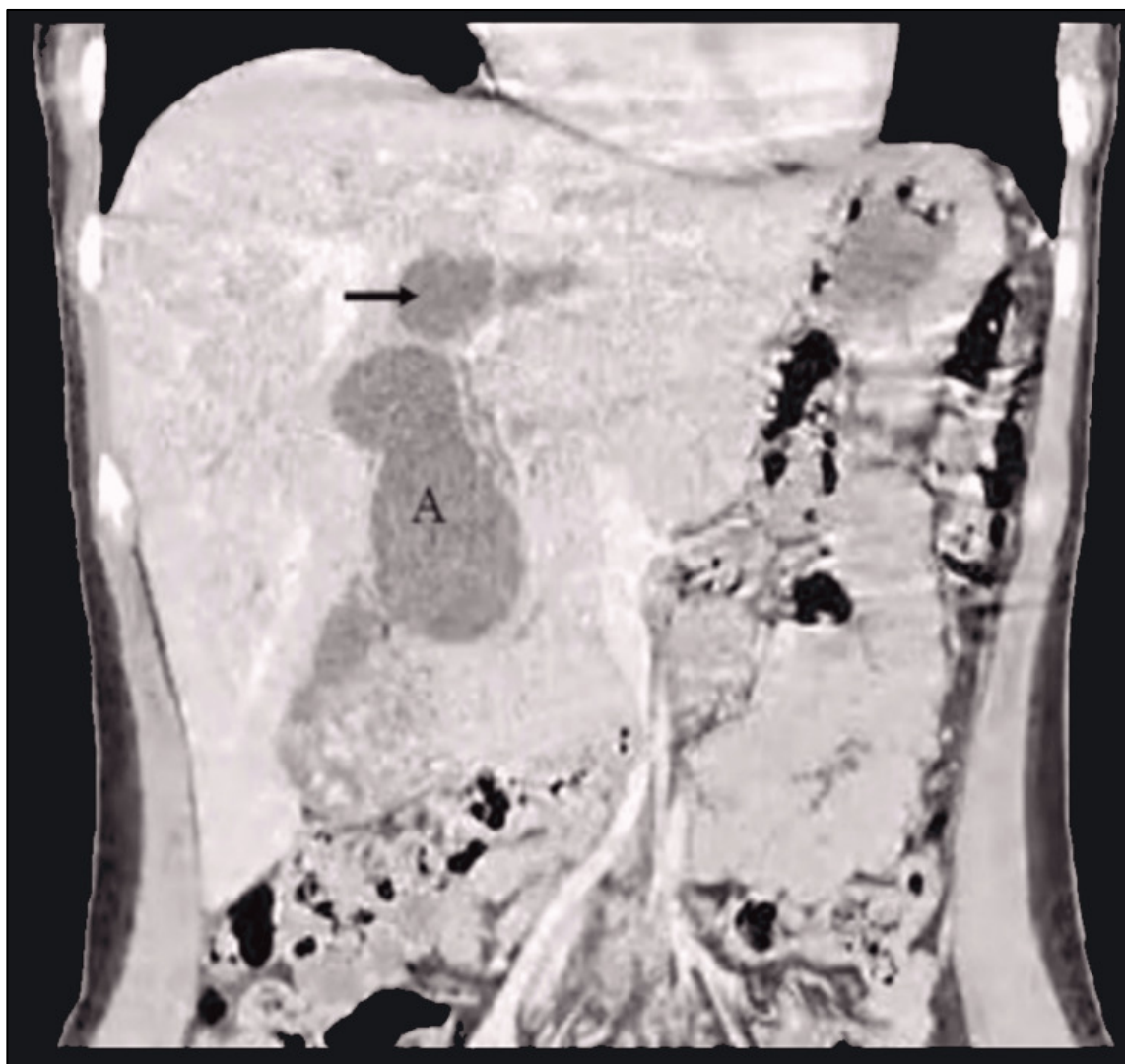


Figure 3. Contrast-enhanced abdominal computed tomography showing intrahepatic and extrahepatic biliary dilatation. Coronal contrast-enhanced abdominal computed tomography showing marked dilatation of the extrahepatic and intrahepatic bile ducts. The dilated common bile duct extends from the supraduodenal portion to the common hepatic duct, with associated dilatation of the right and left hepatic ducts. Reproduced from Villarino Zapata et al. [20].

Table 2. Diagnostic modalities for choledochal cysts.

Modality	Main role	Advantages	Limitations
Ultrasonography	Initial evaluation, pediatric screening, prenatal detection	Accessible, noninvasive, no radiation	Limited distal duct and pancreatobiliary junction assessment [1-3].
MRCP	Preoperative anatomic definition	Noninvasive biliary mapping; defines intrahepatic and extrahepatic anatomy	May require sedation in small children [7,18].
CT	Adult disease, complications, malignancy assessment	Cross-sectional detail; evaluates inflammation, stones, mass, complications	Radiation exposure [19,20].
ERCP	Selected therapeutic or problem-solving indications	Allows sphincterotomy, stone management, sampling, or intervention	Invasive; pancreatitis risk [20,25].
Double-balloon ERCP	Postoperative evaluation in altered anatomy	Access to biliary-enteric reconstruction in selected cases	Requires expertise and specialized equipment [26,31].

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PRINCIPLES OF SURGICAL MANAGEMENT

Definitive treatment for most extrahepatic choledochal cysts consists of complete cyst excision, cholecystectomy, and biliary-enteric reconstruction [1,6-8]. The objectives are to remove abnormal biliary epithelium, eliminate biliary stasis, prevent recurrent cholangitis and pancreatitis, reduce stone formation, preserve hepatic function, and decrease malignancy risk [1,5,6]. Internal drainage alone is no longer considered appropriate definitive therapy because it leaves diseased epithelium in place and is associated with recurrent symptoms and cancer risk [1,6].

Operative planning should consider cyst type, patient age, symptoms, inflammatory status, intrahepatic involvement, hilar anatomy, distal duct anatomy, pancreatobiliary junction anatomy, suspicion for malignancy, and institutional expertise [1,7,8,26]. The standard operation includes careful dissection of the cyst from the portal vein and hepatic artery, distal duct control near the pancreatobiliary junction, cholecystectomy, proximal transection at healthy bile duct, and tension-free biliary reconstruction [7,8,13].

Distal duct management is technically important. Incomplete distal excision may leave a remnant at risk for stones, pancreatitis, recurrent inflammation, or malignancy, whereas overly aggressive dissection can injure the pancreatic duct or pancreatic parenchyma [1,6,33]. Pancreatic leak after cyst resection is uncommon but clinically significant and highlights the need for careful dissection in the intrapancreatic portion of the bile duct [33].

Hilar anatomy also influences outcomes. Hilar duct stenosis, narrow proximal ducts, or type IV-A involvement may require ductoplasty to create a wide, well-vascularized biliary-enteric anastomosis and reduce the risk of postoperative stricture or intrahepatic stone formation [18,29]. In this setting, MRCP is useful for identifying preoperative hilar stenosis and planning reconstruction [18].

Complete surgical excision also provides a specimen for pathologic evaluation, which is particularly important in adults and delayed presentations (Figure 4). Careful pathologic assessment may identify dysplasia, inflammation, fibrosis, or malignancy and helps guide follow-up intensity [5,20,21].

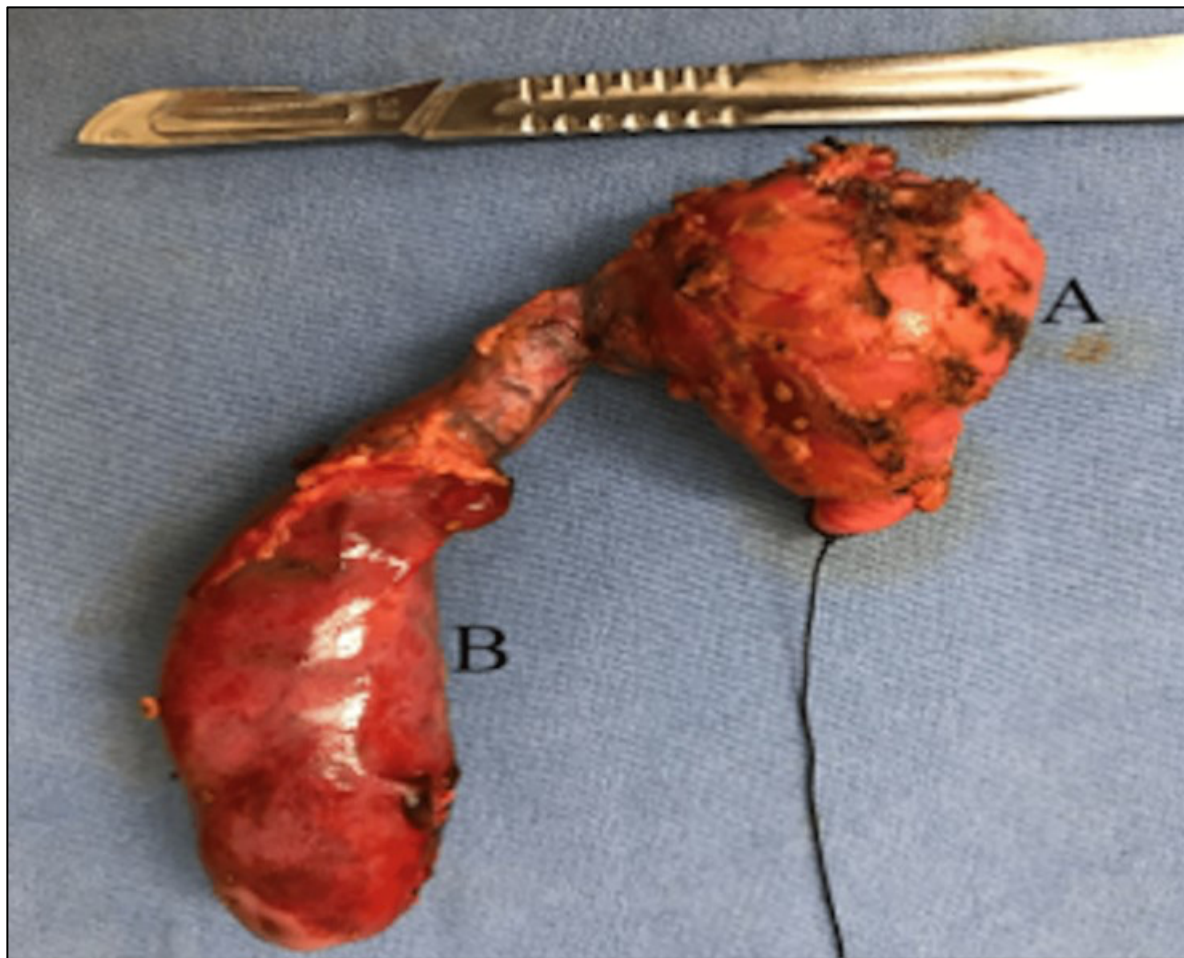


Figure 4. Surgical specimen after choledochal cyst excision and cholecystectomy. Surgical specimen showing en bloc resection of the choledochal cyst and gallbladder. The common bile duct cyst measured 5 cm at its largest diameter, and the gallbladder measured 7 × 3.2 cm. Reproduced from Villarino Zapata et al. [20].

**BILIARY RECONSTRUCTION:
HEPATICOJEJUNOSTOMY
HEPATICODUODENOSTOMY
VERSUS**

After cyst excision, biliary continuity is restored most commonly with Roux-en-Y hepaticojejunostomy or, in selected patients, hepaticoduodenostomy (Figure 5) [10]. Roux-en-Y hepaticojejunostomy has traditionally been considered the standard reconstruction because it provides reliable biliary drainage and reduces direct duodenobiliary reflux [1,7,8,10]. However, it requires creation of a Roux limb and jejunojejunostomy, may be technically demanding

in small children, and can make future endoscopic access more difficult [10,19]. Hepaticoduodenostomy offers technical advantages, including a single anastomosis, shorter reconstruction, and easier endoscopic access to the biliary tree [10]. Potential disadvantages include bile reflux gastritis, duodenobiliary reflux, reflux cholangitis, and theoretical concerns regarding long-term mucosal exposure [10]. Current evidence suggests that both reconstructions may be appropriate in selected patients, but the decision should be individualized according to anatomy, age, surgeon experience, institutional outcomes, and follow-up feasibility [10,12,19].

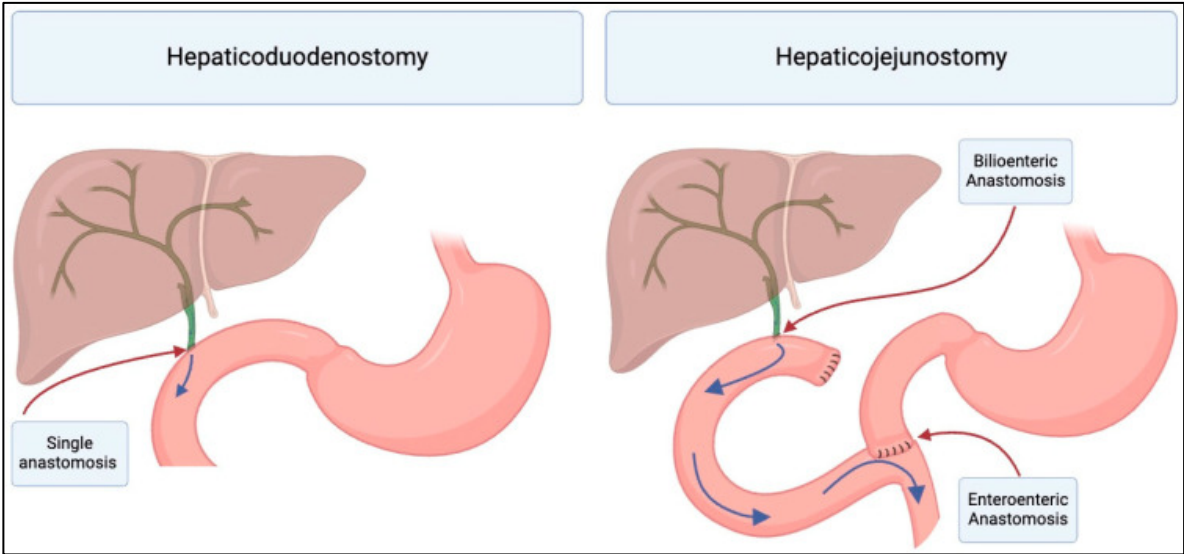


Figure 5. Biliary reconstruction methods after choledochal cyst resection. Schematic illustration of biliary reconstruction after choledochal cyst excision and cholecystectomy. Hepaticoduodenostomy involves a single anastomosis between the proximal bile duct and the duodenum. Hepaticojejunostomy involves creation of a Roux limb, with a biliary-enteric anastomosis to the jejunum and a distal jejunojejunostomy to restore alimentary continuity. Reproduced from Iglesias et al. under the terms of the Creative Commons Attribution 4.0 License [10].

Table 3. Comparison of hepaticojejunostomy and hepaticoduodenostomy after choledochal cyst excision.

Feature	Roux-en-Y hepaticojejunostomy	Hepaticoduodenostomy
Anastomotic configuration	Proximal bile duct to Roux limb	Proximal bile duct to duodenum [1,7].
Main advantage	Established technique; reduced duodenobiliary reflux	Single anastomosis; easier endoscopic access [8,10].
Main limitation	More complex reconstruction; difficult postoperative endoscopic access	Bile reflux, gastritis, reflux cholangitis concerns [10,12].
Technical considerations	Requires Roux limb and jejunojejunostomy	Requires tension-free duct-to-duodenum anastomosis [10,11].
Follow-up implications	Balloon-assisted endoscopy may be needed for access	Conventional endoscopic access may be easier [2,19].

OPEN, LAPAROSCOPIC, AND ROBOTIC APPROACHES

Open surgery remains important in the management of choledochal cysts, particularly in patients with severe inflammation, suspected malignancy, previous biliary surgery, reoperative anatomy, dense adhesions, type IV-A complexity, or limited minimally invasive expertise

[1,6,26,28]. Open surgery provides direct exposure, tactile feedback, and flexibility for complex hilar dissection or oncologic management. In adults, chronic inflammation and adhesions may increase operative difficulty, and malignancy suspicion should prompt careful preoperative planning [5,26,28].

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Laparoscopic excision has become increasingly accepted in experienced centers. Systematic reviews and comparative studies suggest that laparoscopic surgery can be safe and effective in selected children, with potential benefits including reduced surgical trauma, improved cosmesis, faster recovery, and shorter length of stay [9,34-38]. However, laparoscopic cyst excision is technically demanding and requires advanced skills in hepatobiliary dissection and intracorporeal biliary-enteric anastomosis. The quality of the anastomosis is critical because postoperative stricture remains one of the most important long-term complications [12,36].

Enhanced recovery protocols and three-dimensional laparoscopic techniques may further improve perioperative outcomes in selected pediatric patients [37]. Laparoscopic surgery has also been reported as safe and effective in infants in specialized centers, although careful patient selection remains essential [38]. Current practice patterns continue to evolve, and international surveys show variability in the adoption of laparoscopic surgery for choledochal cysts

among pediatric surgeons [39]. Contemporary national data also demonstrate changing trends in surgical excision and outcomes [40].

Robotic surgery has gained increasing attention because it may facilitate precise dissection and intracorporeal suturing, particularly during hepaticojejunostomy [11,13,41-44]. Comparative and systematic review data suggest that robotic-assisted cyst excision can achieve favorable outcomes in selected pediatric patients, although operative time, cost, availability, and surgeon experience remain important limitations [11,13,41,42]. Robotic techniques have been described in neonates and patients weighing 10 kg or less, reflecting expanding feasibility in specialized centers [43,44]. In adults, laparoscopic and robotic surgery for congenital biliary dilatation can achieve favorable short-term outcomes when performed by experienced hepatobiliary teams [28].

Recent comparative experience continues to evaluate laparoscopic and robotic approaches in pediatric choledochal cyst surgery [41,47].

Table 4. Surgical approaches for choledochal cyst excision.

Approach	Advantages	Limitations	Best suited for
Open surgery	Direct exposure; useful in complex anatomy; appropriate when malignancy is suspected	Larger incision; longer recovery; more surgical trauma	Severe inflammation, adults with complicated disease, suspected cancer, reoperations [1,6].
Laparoscopic surgery	Less invasive; improved cosmesis; faster recovery in selected cases	Advanced learning curve; demanding biliary anastomosis	Experienced centers, selected pediatric and adult patients [9,11,28].
Robotic surgery	Improved ergonomics; precise suturing; useful for reconstruction	Cost, availability, operative time, specialized training	Specialized centers, selected pediatric, neonatal, low-weight, and adult patients [34-44].

SPECIAL POPULATIONS

Prenatally Diagnosed Neonates

Prenatal diagnosis has increased with widespread obstetric ultrasonography. Prenatally detected cystic biliary lesions require postnatal confirmation, liver function assessment, evaluation for biliary obstruction, and planning of surgical timing [25,26]. Neonatal surgery may be indicated in symptomatic patients, progressive biliary obstruction, suspected perforation, worsening liver dysfunction, or rapid cyst enlargement. However, early surgery must be balanced against the technical challenges of small anatomy, fragile tissues, and narrow bile ducts [25,26].

Recent studies suggest that minimally invasive and robotic approaches may be feasible even in selected neonates and

low-weight children when performed by experienced teams [25,43,44]. Nevertheless, these cases require specialized expertise and careful perioperative planning.

Infants and Children

Children remain the most common population treated for choledochal cysts. Early definitive excision aims to prevent recurrent cholangitis, pancreatitis, progressive fibrosis, and later malignancy risk [7,8,27,32]. Minimally invasive approaches are increasingly used in children, but outcomes depend on surgeon experience, anastomotic quality, and appropriate patient selection [9,34-44].

Adults

Adults with choledochal cysts often present after years of chronic inflammation, biliary symptoms, pancreatitis, stones,

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or incidental imaging findings [1,5,24,26,28]. Operative management may be more difficult because of adhesions, inflammation, distorted anatomy, and malignancy concern. Complete cyst excision remains the goal when feasible, but adult patients require careful preoperative evaluation for cholangiocarcinoma and gallbladder carcinoma [5,21]. Minimally invasive and robotic approaches may be considered in selected adult patients in experienced centers [28].

POSTOPERATIVE COMPLICATIONS

Postoperative complications can be early or late. Early complications include bile leak, bleeding, pancreatitis, pancreatic leak, intra-abdominal collections, ileus, wound complications, and anastomotic problems [1,7,8,33,34]. Pancreatic leak may occur when distal dissection is difficult or when the intrapancreatic duct is closely associated with the pancreatic duct [33].

Late complications are particularly important because they may occur years after surgery. Anastomotic stricture is one of the most significant long-term complications, as it can lead to cholangitis, intrahepatic ductal dilatation, hepatolithiasis, secondary biliary cirrhosis, and need for reintervention [12,14-19]. A systematic review and meta-analysis focusing on postoperative anastomotic stricture highlights the need for careful surgical technique and long-term monitoring [12].

Intrahepatic bile duct stones are another clinically relevant late complication, especially in type IV-A disease, hilar stenosis, or impaired biliary drainage [17,18]. Management may require endoscopic, percutaneous, or surgical intervention. In patients with altered anatomy after biliary-enteric reconstruction, double-balloon endoscopic retrograde cholangiography may provide diagnostic and therapeutic access in selected cases [19]. Long-term studies show that complications may occur within the first several years after surgery but can also develop later, supporting structured and prolonged surveillance [14-16].

Table 5. Early and late complications after choledochal cyst excision.

Complication	Timing	Diagnostic approach	Management
Bile leak	Early	Drain output, bilirubin level, ultrasound or CT	Drainage, supportive care, reintervention if persistent [12,14].
Pancreatitis or pancreatic leak	Early	Serum enzymes, drain amylase, CT	Supportive care, drainage, specialized pancreatic management [15,16].
Anastomotic stricture	Late	Liver tests, ultrasound, MRCP, cholangiography	Endoscopic, percutaneous, or surgical treatment [16,17].
Intrahepatic stones	Late	Ultrasound, MRCP, cholangiography	Endoscopic/percutaneous extraction, ductoplasty, or surgery [15,18].
Cholangitis	Early or late	Clinical findings, inflammatory markers, imaging	Antibiotics, biliary drainage, stricture correction [18,19].
Malignancy	Late	Imaging, tumor markers, pathology	Hepatobiliary oncology evaluation [19,33].

LONG-TERM OUTCOMES AND SURVEILLANCE

Successful cyst excision does not eliminate the need for long-term follow-up. Patients remain at risk for anastomotic stricture, cholangitis, intrahepatic stones, pancreatitis, liver dysfunction, and malignancy [12-21]. Surveillance should be individualized based on cyst type, age at surgery, reconstruction method, operative findings, intrahepatic involvement, postoperative complications, and pathology. Routine follow-up generally includes clinical assessment and liver function testing. Ultrasonography is useful as a noninvasive screening tool for biliary dilatation, stones, and

liver changes. MRCP should be considered when patients develop abdominal pain, cholangitis, abnormal liver tests, suspected stricture, suspected intrahepatic stones, or unclear postoperative anatomy [18,19]. In patients with Roux-en-Y anatomy, access to the biliary tree may be challenging, and double-balloon enteroscopy-assisted cholangiography may be useful in selected postoperative complications [19].

Long-term outcome studies emphasize that complications may occur even after technically successful surgery. Late postoperative complications in pediatric patients and single-center long-term experiences demonstrate the importance of

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structured surveillance, particularly within the first five years after surgery and beyond [14-16]. Patients with type IV-A disease, hilar stenosis, intrahepatic duct abnormalities, recurrent cholangitis, or intrahepatic stones require closer follow-up [17,18,29].

Long-term follow-up remains important after pediatric choledochal cyst excision, as late complications and loss of surveillance may affect postoperative care [14-16,46].

Table 6. Suggested long-term surveillance strategy after choledochal cyst excision [14-19].

Step	Evaluation	Purpose
1	Clinical assessment	Identify abdominal pain, fever, jaundice, cholangitis, pancreatitis symptoms
2	Liver function tests	Detect cholestasis, hepatocellular injury, or progressive dysfunction
3	Ultrasonography	Screen for biliary dilatation, intrahepatic stones, liver changes
4	MRCP if symptoms or abnormal tests	Define anatomy, stricture, stones, intrahepatic disease
5	Endoscopic/percutaneous/surgical referral	Treat stricture, stones, cholangitis, or complex complications
6	Lifelong surveillance in selected patients	Monitor malignancy risk and late complications

MALIGNANCY RISK

Malignancy risk is one of the central reasons for definitive treatment and long-term follow-up. Choledochal cysts are associated with an increased risk of biliary tract cancer, including cholangiocarcinoma and gallbladder carcinoma [1,5,6,21]. Risk appears to increase with age, delayed diagnosis, chronic inflammation, pancreatobiliary reflux, incomplete excision, and persistent abnormal biliary epithelium [5,21]. Complete excision reduces but does not completely eliminate cancer risk. Residual risk may persist due to intrahepatic duct involvement, longstanding inflammation, incomplete resection, or field changes in the biliary epithelium [1,5,21]. Molecular studies support the concept that carcinogenesis in congenital biliary dilatation may involve genetic and epigenetic regulation, reinforcing the importance of early definitive treatment and long-term surveillance [21-23]. Adults require particular attention because malignancy rates are higher in delayed presentations. A systematic review of adult choledochal cysts diagnosed in Western populations emphasized the importance of malignancy assessment in this group [5]. Suspicious symptoms or imaging findings should prompt comprehensive evaluation, including high-quality cross-sectional imaging, MRCP, laboratory testing, possible tumor marker assessment, and referral to hepatobiliary oncology when indicated [1,5,21].

CURRENT CONTROVERSIES AND FUTURE DIRECTIONS

Several controversies remain. First, the optimal biliary reconstruction continues to be debated. Roux-en-Y hepaticojejunostomy remains widely used, but hepaticoduodenostomy may be appropriate in selected patients because of its simplicity and endoscopic accessibility [10]. Long-term comparative data are needed to better define patient selection, reflux-related complications, quality of life, and reintervention rates. Second, the role of minimally invasive and robotic surgery continues to evolve. Laparoscopic surgery has demonstrated favorable outcomes in selected patients, but results depend heavily on surgeon experience and anastomotic quality [9,34,38]. Robotic surgery may facilitate precise suturing and complex reconstruction, but cost, availability, operative time, and long-term outcomes remain important considerations [11,13,41-44]. Robotic surgery in neonates and low-weight children is promising but should remain concentrated in centers with specialized expertise until broader evidence is available [43,44]. Third, the timing of surgery in prenatally diagnosed neonates remains individualized. Early surgery may prevent complications, but neonatal operation is technically challenging. Decisions should consider symptoms, cyst size, liver function, biliary obstruction, suspected perforation, and institutional expertise [25,26].

Choledochal Cysts: Contemporary Diagnosis, Surgical Management, and Long-Term Surveillance

Future directions include standardized surveillance protocols, improved risk stratification for malignancy, integration of molecular biomarkers, multicenter comparative studies of reconstruction techniques, and better definition of the role of robotic surgery [16,21,23,45]. Bibliometric analysis shows growing global interest in the surgical management of choledochal cysts, reflecting the need for continued international collaboration and long-term outcome research [45].

LIMITATIONS

This review has several limitations. First, it was designed as a narrative review rather than a systematic review or meta-analysis; therefore, study selection was qualitative and may not include all available evidence. Second, the literature on choledochal cysts is heterogeneous, with substantial variation in patient age, cyst type, operative approach, reconstruction technique, follow-up duration, and outcome reporting. Third, many contemporary studies are retrospective and originate from high-volume specialized centers, which may limit generalizability to institutions with lower case volume or limited minimally invasive and robotic expertise. Despite these limitations, this review synthesizes recent clinically relevant evidence and provides a practical surgical framework for diagnosis, operative management, and long-term surveillance.

CONCLUSIONS

Choledochal cysts remain clinically important congenital biliary abnormalities with implications ranging from prenatal diagnosis to adult hepatobiliary oncology. Contemporary management requires accurate anatomic diagnosis, careful classification, individualized operative planning, complete cyst excision when appropriate, cholecystectomy, and durable biliary reconstruction. MRCP plays a central role in preoperative evaluation, whereas ultrasonography, CT, and ERCP remain useful in selected clinical contexts. Laparoscopic and robotic approaches are increasingly feasible in specialized centers, but outcomes depend on appropriate patient selection, technical expertise, and anastomotic quality. Long-term follow-up is essential because postoperative strictures, intrahepatic stones, cholangitis, liver dysfunction, and residual malignancy risk may occur even after technically successful surgery. Future progress will depend on standardized surveillance protocols, multicenter outcome data, and improved risk stratification for late complications and malignancy.

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