Diagnosis and Treatment of Choledochoceles

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Choledochoceles are cystic dilatations of the intraduodenal portion of the common bile duct. Although often classified as Type III biliary cysts, choledochoceles have distinctive demographic and anatomic features and a lower risk of malignancy than other types of choledochal cysts. Type A choledochoceles are cystic dilatations of a segment of the intra-ampullary bile duct and are located proximal to the ampullary orifice. Type B choledochoceles are diverticula of the intra-ampullary common channel and are located distal to the ampullary orifice; they can be distinguished from duodenal duplication cysts both anatomically and histologically. Both types of choledochocele may present with pancreatitis, biliary obstruction, or nonspecific gastrointestinal symptoms. Cross-sectional imaging, endoscopic ultrasound, and endoscopic retrograde cholangiopancreatography are useful for diagnosis. Choledochoceles may be drained or resected endoscopically. Surveillance for dysplasia should be considered for lesions that are not resected.

Keywords: Choledochal Cyst; Sphincterotomy.

The Greek prefix choledocho- refers to the common bile duct (CBD), and the suffix cele refers to a swelling or cavity. The term choledochocele was coined by Wheeler in 1940 to describe a cystically dilated intraduodenal portion of the CBD. Choledochoceles are often categorized as a subtype of choledochal cysts, a heterogeneous group of anomalous dilatations of the biliary tree first described by Abraham Vater and generally considered congenital in origin. Choledochal cysts are usually organized into 5 subgroups that are based on characteristic findings: Type I, a spherical or fusiform dilation of the CBD; Type II, a single diverticulum of the CBD; Type III, a cystic dilatation of the distal CBD within the ampulla of Vater and protruding into the duodenum (also called choledochocele); Type IV, multiple dilatations involving the intrahepatic and/or extrahepatic biliary tree; and Type V, intrahepatic bile duct cysts (also known as Caroli’s disease). This review focuses on Type III choledochal cysts or choledochoceles, as well as duodenal duplication cysts, which overlap with choledochoceles in the medical literature.

Epidemiology and Etiology

Choledochal cysts are uncommon, and the prevalence may vary by region, because they are reported disproportionately from Asia. Choledochoceles, or Type III choledochal cysts, have been considered the least common subtype, representing <5% of all reported cysts; however, a recent study from a Western institution reported that almost 20% of their cohort of 146 choledochal cysts were choledochoceles. The true incidence of choledochoceles is likely dependent on the definition of the lesion, the diagnostic modalities available, and the population studied.

Choledochoceles frequently present at an older age when compared with other choledochal cysts, with an average age at presentation of 51 years compared with 29 years. Pediatric presentations are unusual for choledochoceles but common for other types of choledochal cysts. Typically, a female predominance is observed in choledochal cysts, but this gender predilection is not apparent in persons with choledochoceles. In contrast to other choledochal cysts, the risk of malignancy appears to be lower in choledochoceles. These considerations have led some to conclude that choledochoceles should not be classified as choledochal cysts.

It is likely that most choledochal cysts arise in utero or early infancy; however, the origin of choledochoceles has been long debated. Schwegler and Boyden postulated that the primitive ampulla arises from 2 ostia during embryonic development, with the inferior ostium subsequently regressing and the superior ostium becoming the ampulla of Vater. By using this paradigm, a choledochocele could develop from a rudimentary bile duct that, after failed or transposed regression, may grow larger in size. Additional theories hypothesize that these lesions represent a congenital cyst or diverticulum of the intramural segment of the CBD. Choledochoceles might also be acquired, possibly as a result of papillary inflammation leading to obstructive ballooning of the intramural CBD or a secondary effect of sphincter of Oddi dysfunction or stenosis, with the increased sphincter pressure causing development of

Abbreviations used in this paper: CBD, common bile duct; CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography; EUS, endoscopic ultrasound; MRCP, magnetic resonance cholangiopancreatography; MRI, magnetic resonance imaging; US, ultrasound.
a cystic dilatation.\textsuperscript{17} Because of the variety of proposed pathogeneses and variance of age at presentation, it seems likely that both congenital and acquired forms of choledochocele probably exist.

**Classification**

Cyst anatomy, histology, and radiology have all been used to classify periampullary cystic lesions, with various terminologies resulting. Currently there is no uniform agreement regarding their classification. Sarris and Tsang\textsuperscript{6} subdivided choledochoceles into Type A and Type B lesions on the basis of their anatomic appearance. Type A lesions are those in which the intramural bile duct opens into a cystically dilated segment, which communicates to the duodenal lumen via a separate orifice (Figures 1A and 2). Type B choledochoceles are characterized by a bile duct that opens normally into the duodenal lumen, with the choledochocele arising as a diverticulum of the intra-ampullary common channel (Figures 1B and 3). Type B choledochoceles are often lined by duodenal mucosa. Additional choledochocele variants (Types C–E) have been described by Kagiyama et al.,\textsuperscript{17} with each variant demonstrating slightly different pancreaticobiliary anatomy.

Type B choledochoceles, as described by Sarris and Tsang,\textsuperscript{6} have also been referred to as duodenal duplications\textsuperscript{18} and diverticular choledochoceles.\textsuperscript{15,19} Antaki et al.\textsuperscript{18} narrowly defined choledochoceles as cystic lesions lined with biliary mucosa arising from the intramural CBD and appearing proximal to the papilla when visualized endoscopically. In contrast, duodenal duplications were defined as fluid-filled structures lined by duodenal mucosa that protrude into the duodenal lumen distal to the papilla.\textsuperscript{19} However, both Type A and Type B choledochoceles may be lined by either duodenal or biliary mucosa.\textsuperscript{6,20,21} Radiologic criteria differentiating choledochoceles from duodenal duplications have also been proposed,\textsuperscript{15} which delineate these lesions on the basis of cholangiography and upper gastrointestinal barium studies. By using these criteria, choledochoceles fill with contrast during cholangiography but not during barium studies, whereas the lack of filling during both studies along with the absence of bile content within the cyst defines duodenal duplications.

A choledochocele is defined by its relationship to the bile duct, whereas duodenal duplications are defined by their relationship to the duodenum. Duplications may occur throughout the alimentary tract. They typically arise from the mesenteric border of the gut, share a common wall with the intestine, and may or may not communicate with the gut lumen. Histologically they have a well-defined smooth muscle coat and often have redundant smooth muscle layers.\textsuperscript{22–24} The mucosal lining of a duplication is not indicative of its site of origin; ileocolonic duplications, for instance, may be lined by gastric or respiratory mucosa.\textsuperscript{25–27} This suggests that mucosal histology should not determine classification.

We propose that cystic lesions that arise from the intramural bile duct or intra-ampullary common channel, with direct anatomic communication to these ducts, be considered choledochoceles. These may be lined by biliary or enteric mucosa but do not fuse with the duodenal wall and do not have a muscle layer other than muscularis mucosa. Multiple variants exist,\textsuperscript{17} but the most common are Types A and B as defined by Sarris and Tsang\textsuperscript{6} (Figure 1A and B). Choledochoceles lined with biliary mucosa may be acquired because of the above-mentioned mechanisms, whereas those lined with duodenal mucosa might occur as a result of a congenital foregut anomaly. Duodenal duplications (Figure 1C) are also cystic structures typically found in the second or third portions of duodenum, but in contrast to choledochoceles, they are intimately attached to the duodenal wall, sharing a portion of their circumference with the duodenum. They have a muscle coat, often with redundant smooth muscle layers.\textsuperscript{28–30} Almost 50% of duodenal duplications communicate with the peri-ampullary ducts, often to the pancreatic duct, and at times via an aberrant duct.\textsuperscript{31}

![Figure 1. Anatomy of choledochoceles and duodenal duplication cysts. (A) Type A choledochocele. (B) Type B (diverticular) choledochocele. (C) Duodenal duplication cyst.](image-url)
Intraluminal duodenal diverticulum, also called “windsock” diverticulum, is another congenital duodenal anomaly. This lesion, which is characterized by an intraluminal mucosal diaphragm that forms an intraluminal diverticulum, is found adjacent to the papilla but does not communicate with the bile duct. The diagnosis and treatment of intraluminal duodenal diverticulum have been recently reviewed.32

**Clinical Presentation**

The initial clinical symptoms of choledochoceles vary widely and are nonspecific. Upper abdominal pain with concomitant nausea and vomiting is present in more than 90% of symptomatic cases.9 Physical exam findings are equally nonspecific and generally modest but may reveal abdominal distention, tenderness to palpation, or palpable mass in rare cases.31 Choledochoceles may be identified incidentally in patients undergoing radiologic evaluation for unrelated reasons.

As shown in Table 1, the presentation of choledochoceles differs from that of other types of biliary cysts. Pancreatitis is the most common clinical complication associated with symptomatic choledochoceles and is frequently the inciting event that leads to identification of the choledochocele.5,9,33 Patients often have acute relapsing pancreatitis with multiple discrete episodes.18 The development of acute pancreatitis in this setting has been attributed to obstruction of pancreaticobiliary outflow leading to increased ductal pressures, or bile reflux into the pancreatic duct.34 External compression of the papilla leading to pancreatitis has been described in cysts that enlarge because of accumulation of stones or debris9,35 and in cases where the sheer mass of the lesion is sufficient to obstruct pancreaticobiliary drainage.36 Stones form within the cystic dilation in about 20% of cases, likely because of chronic stasis.6 Jaundice and cholangitis are less common presentations when compared with other types of choledochal cysts.5,33

Multiple unusual clinical presentations have also been described in conjunction with choledochoceles. Cases with associated pancreas divisum,25 intestinal intussusception,17 gastric outlet obstruction,38 gastrointestinal bleeding,39 and primary sclerosing cholangitis10 have been described. In addition, duplication cysts of the duodenum have been associated with other congenital gastrointestinal malformations including imperforate anus, intestinal malrotation, biliary atresia, and double gallbladder.12,41

**Risk of Malignancy**

Choledochoceles and duodenal duplication cysts are benign, but malignancy within these lesions has been described. The incidence of carcinoma in patients...
with symptomatic choledochoceles has been estimated at 2.5%,\textsuperscript{10,11} and only a handful of cases with concurrent ampullary carcinoma or cholangiocarcinoma have been reported.\textsuperscript{5} Malignancy may develop in both Type A and Type B choledochoceles. In one case, dysplasia and subsequent carcinoma were detected during follow-up endoscopic surveillance of a patient with a Type A choledochoele that had been previously treated with endoscopic sphincterotomy.\textsuperscript{10} Pancreatic cancer also rarely occurs in association with choledochoceles.\textsuperscript{5,42,43}

The apparently low incidence of associated malignancy is in stark contrast to other types of choledocho cysts, which carry a substantial risk of malignancy, both gallbladder carcinoma and cholangiocarcinoma.\textsuperscript{31,44} This increased risk is at least in part attributable to the presence of an anomalous pancreaticobiliary junction, which is frequently seen in patients with Types I and IV choledochal cysts.\textsuperscript{45} Malignancy in this setting may arise from the reflux of pancreatic juice into the biliary tree, with subsequent pancreatic enzyme activation leading to chronic mucosal inflammation and culminating in biliary dysplasia and carcinoma.\textsuperscript{34,46} Pancreaticobiliary junction abnormalities are less common in patients with choledochoceles, occurring in <20% of patients, perhaps accounting for the decreased risk of malignancy compared with other choledochal cysts;\textsuperscript{5} however, similar mechanisms of pancreaticobiliary reflux or stasis may occur and contribute to malignant development.

The risk of malignancy in patients with duodenal duplications also appears to be low. To date, only several case reports have described concurrent malignancy in patients with duodenal duplication cysts,\textsuperscript{47–49} including a carcinoid tumor and 2 cases of adenocarcinoma occurring within the duplication. Overall, malignant change within all gastrointestinal tract duplications is quite uncommon, with only 30 reported cases as of 2006.\textsuperscript{48} The pathogenesis of cancer in this setting remains poorly defined but may also be related to chronic mucosal inflammation and subsequent metaplasia.\textsuperscript{48}

**Diagnosis**

Before the advent of modern imaging technologies, choledochoceles were identified during open surgical exploration\textsuperscript{1,50} or by intravenous cholangiography.\textsuperscript{15} In the modern era, patients are often evaluated by using more than one imaging modality, with each study providing complementary information. Plain abdominal radiographs are generally unhelpful, but gastrointestinal contrast studies can reveal smooth submucosal lesions, ovoid filling defects, or extrinsic masses extending within the duodenum.\textsuperscript{51} Barium studies may be sufficient to identify larger anomalies, but smaller lesions may be overlooked. The combination of an intraluminal duodenal filling defect on barium study that also opacifies on cholangiography is suggestive of a choledochoele.\textsuperscript{6}

Conventional abdominal ultrasound (US) is frequently the initial diagnostic test performed in patients undergoing upper abdominal evaluation, but the accuracy for detection of choledochoceles is poor (31%), which is likely related to the inherent difficulty of evaluating the duodenum and distal CBD by using this modality.\textsuperscript{1} When visible on US, choledochoceles often have a thick wall, and calculi may be visible within the cyst.\textsuperscript{52} Duodenal duplications can be distinguished sonographically from choledochoceles by their pattern of wall layers, with a hypoechoic layer in the cyst wall representing smooth muscle.\textsuperscript{53} Peristalsis of a duodenal duplication may also be observed on conventional US.\textsuperscript{54}

Cross-sectional imaging with computed tomography (CT) or magnetic resonance imaging (MRI) is more accurate than US and provides more specific information related to the location, size, and anatomic relationship to the pancreaticobiliary tree.\textsuperscript{33} In addition, these studies are less invasive than upper endoscopy, endoscopic US (EUS), and endoscopic retrograde cholangiopancreatography (ERCP) and can provide evidence of associated intra-abdominal pathology such as acute pancreatitis and biliary obstruction.\textsuperscript{54,55} CT and MRI are capable of identifying small lesions missed on conventional US or barium studies. Classically, cross-sectional imaging demonstrates a distinct cystic structure on the medial wall of the descending duodenum with central fluid density or inhomogeneity due to biliary sludge and calculi.\textsuperscript{9,33} Cyst wall calcification may also be observed.\textsuperscript{51} Magnetic resonance cholangiopancreatography (MRCP) and CT cholangiography may further enhance the diagnostic capabilities of standard CT and MRI. CT cholangiography, which uses intravenous contrast medium that is excreted into the biliary tree, currently offers higher resolution than MRCP but exposes the patient to

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**Table 1. Clinical Presentation of Choledochoceles, Other Choledochal Cysts, and Duodenal Duplications**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Choledochoceles (%)</th>
<th>Other choledochal cysts (Types I, II, IV, V) (%)</th>
<th>Duodenal duplications (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pancreatitis</td>
<td>38–70</td>
<td>0–31</td>
<td>50</td>
</tr>
<tr>
<td>Jaundice</td>
<td>11–25</td>
<td>30–73</td>
<td>3</td>
</tr>
<tr>
<td>Cholangitis</td>
<td>0–10</td>
<td>21–70</td>
<td>3</td>
</tr>
</tbody>
</table>

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\textsuperscript{44,46,68}
radiation. Both MRCP and CT cholangiography images can be reconstructed by using three-dimensional techniques, providing further definition and clarity of the pancreaticobiliary anatomy.

Cross-sectional imaging is frequently followed by endoscopic evaluation. Choledochoceles have a characteristic endoscopic appearance, with Type A lesions presenting as a rounded protrusion of the intramural CBD proximal to the ampullary orifice (Figure 2) and Type B lesions appearing as pendulous masses in the duodenal lumen, inferior to the ampullary orifice (Figure 3). The overlying mucosal surface is generally smooth and soft to palpation, frequently resembling a submucosal tumor. They may be seen with standard forward-viewing endoscopes but are better visualized by using side-viewing endoscopes. EUS may demonstrate communication with the pancreaticobiliary tree. Similar to transabdominal US, EUS shows the wall layers of the cyst as well as stones, sludge, or mucosal masses within it. In addition, EUS can demonstrate whether the lesion is separate from the duodenal muscle layers (choledochocele) or in continuity with them (duodenal duplication), guiding both classification of the lesion and treatment decisions.

ERCP remains the most widely used diagnostic tool for identification of choledochoceles, with a reported diagnostic sensitivity of 97%. ERCP also offers the potential for treatment of the lesion during the same endoscopic procedure. Enlargement of a Type A choledochocele occurs as contrast is injected into the bile duct during ERCP, and the diagnosis may not be apparent until the lesion balloons in this way. Cholangiography shows retention of contrast medium within the cystic dilation at the terminus of the CBD. Contrast filling at ERCP usually occurs in Type B (diverticular) choledochoceles and may or may not occur in duodenal duplication cysts, depending on the anatomic relationship of the cyst to the pancreaticobiliary tree and the cannulation method used.

### Treatment

Choledochoceles may be treated by resection or drainage into the gut lumen. For other types of choledochal cysts, resection is favored over drainage because of the high risk of malignancy, but the low risk of malignancy in choledochoceles makes drainage a reasonable treatment option for many patients. The choice of treatment modality depends on the patient’s age, symptoms, and comorbid conditions, as well as the size of the lesion and its anatomic relationship to the ducts and duodenal wall. For instance, in a young patient, complete excision of a Type B choledochocele may be desirable in part because of the subsequent risk of dysplasia, whereas in an older patient with a Type A choledochocele, endoscopic drainage of the lesion (by sphincterotomy) is probably sufficient.

Historically, surgical intervention was considered the standard of care to relieve ductal obstruction and excise potentially premalignant tissue; however, surgery can be complex because of the close proximity of the lesions to the pancreaticobiliary confluence. The most commonly performed surgical procedure is transduodenal excision, with or without sphincteroplasty. Sarris and Tsang proposed that Type A choledochoceles be treated with local excision with ampullary preservation, whereas Type B lesions required both excision and sphincteroplasty. Other surgical procedures less commonly performed include surgical unroofing (or marsupialization) with sphincteroplasty, cyst excision with hepaticojejunostomy, or pancreaticoduodenectomy.

Dehyle and Meyer first described endoscopic management of a choledochocele in 1974. This was followed by reports from Siegel et al describing endoscopic sphincterotomy in patients with Type A choledochoceles. Endoscopic therapy is now widely considered the standard of care in most patients with symptomatic choledochoceles (Figure 2). Because they are located proximal to the ampullary orifice and in continuity with the bile duct, Type A choledochoceles are typically treated by endoscopic sphincterotomy. A novel technique that uses endoscopic snare resection of Type A choledochoceles with balloon-catheter assistance has also been described by using either single-channel or double-channel endoscopes. In this technique a standard polypectomy snare is passed over a deeply placed biliary wire during ERCP. A balloon catheter is then also passed over the guidewire and through the snare into the choledochocele, where it is inflated and withdrawn toward the endoscope, tenting the choledochocele into the lumen. The snare is then closed around the base of the choledochocele, and the tissue is excised. This technique, which is akin to surgical marsupialization, removes a major portion of the choledochocele wall. In addition, standard snare resection of Type A choledochoceles, without balloon-catheter assistance, may be performed successfully.

Type B choledochoceles, which are distal to the ampullary orifice, have been treated endoscopically by a number of methods. These include endoscopic incision of the luminal aspect of the cyst, with or without balloon dilation or stent placement, as well as marsupialization by partial snare excision of the cyst wall, or complete snare resection. Complete resection most closely parallels the surgical approach to these lesions (Figure 3) and is desirable because it removes the lesion, obviating concerns about subsequent endoscopic surveillance for dysplasia. To encompass the entire lesion with a snare, the cyst may have to be deflated by incision or aspiration before resection.

Because duodenal duplication cysts (as we define them) share muscle layers with the duodenal wall, they have not been completely resected endoscopically. Nevertheless, endoscopic drainage is an important management option for duodenal duplication cysts. This can
be accomplished by needle-knife incision or snare resection of the luminal aspect of the lesion, which unroofs or marsupializes the cyst cavity.\textsuperscript{118,67} Duodenal duplications can also be resected surgically and may require partial duodenectomy or pancreaticoduodenectomy.\textsuperscript{10,30}

We believe that most Type A choledochoceles should be treated by using either endoscopic sphincterotomy or snare resection, whereas most Type B choledochoceles should be completely removed endoscopically by using a polypectomy snare. Although endoscopic intervention may be sufficient in most cases, lesions whose anatomy cannot be defined or varies significantly should be considered for surgery, as well as those that harbor malignancy. Duodenal duplications will more often undergo surgical resection, particularly in young patients. Lesions treated with a drainage procedure rather than resection should be biopsied for histology.

**Prognosis and Follow-up**

Few studies have evaluated the outcomes of patients after treatment, but the available evidence suggests that either surgical or endoscopic intervention leads to rapid symptom resolution, with few reported subsequent complications.\textsuperscript{5,10,18} As previously mentioned, there is a poorly defined but apparently low risk of malignancy arising in choledochoceles and duodenal duplication cysts, and malignancy may become apparent after prior treatment. When these lesions are not completely resected, the pros and cons of surveillance (including periodic endoscopy and mucosal biopsies within the lesion) should be discussed with the patient. The best interval and overall duration of endoscopic surveillance are unknown, but some have proposed repeat evaluation 6–12 months after the index endoscopic procedure.\textsuperscript{18}

**Summary**

Choledochoceles are typically diagnosed and managed by gastroenterologists. The diagnosis is often first entertained when CT or MRI demonstrates a cystic lesion in the duodenum. The diagnosis is further clarified by EUS and ERCP, which will distinguish Type A and Type B choledochoceles from duodenal duplication cysts and guide management decisions. Drainage or resection of choledochoceles can be accomplished endoscopically. Although the risk of malignancy is low compared with other types of biliary cysts, choledochoceles should be evaluated histologically. Consideration should be given to post-treatment surveillance of choledochoceles that are managed by sphincterotomy or other forms of endoscopic drainage, particularly in young patients.

**References**


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Conflicts of interest
The authors disclose no conflicts.

1. The most common complication of a choledococele is:
   a. biliary sepsis
   b. painless jaundice
   c. acute pancreatitis
   d. duodenal obstruction

**True or False**

2. Choledococeles are type III choledocal cysts, affecting the distal CBD and associated with a lower malignancy rate compared to other choledocal cysts

3. Choledococeles present most often in older individuals; most often females

4. Choledochoceles may be treated with endoscopic resection and do not always require surgery

5. Duodenal duplication cysts should not be removed endoscopically, but drainage using a needle-knife incision or snare unroofing of the lesion should be done.

6. Duodenal duplication cysts do not fill with contrast during ERCP

7. The incidence of malignancy in patients with symptomatic choledochoceles is 2.5%

8. Endoscopically, choledochoceles resemble submucosal tumors of the duodenum

9. Type B choledococele is located proximal to the ampullary orifice

10. After endoscopic therapy of a choledochocele, follow up endoscopy with biopsy is recommended in 6-12 months