Biliary Cysts

A Review and Simplified Classification Scheme

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KEYWORDS

• Biliary cysts • Review • Classification scheme • Simplified

KEY POINTS

- Various biliary cystic conditions that have been considered together really represent markedly different disorders in terms of embryologic and anatomic considerations, and they also carry different risks of both neoplastic and nonneoplastic complications.
- Modern imaging techniques, as well as advances in operative technique and our ability to manage patients in the perioperative period, have likely altered and simplified the necessary classification schemes and treatment algorithms.

INTRODUCTION

If one has no real knowledge of biliary cysts, then one should study them. After a short time, it becomes clear that much is known about cystic abnormalities of the biliary system. If one continues to study theses oddities, perhaps for a long time, then it becomes even clearer that we do not know much at all about biliary cysts. There are competing views of how they come to be. There are competing views of how to classify them. There are even competing views of whether or not they are cysts.[1]

Despite the lack of agreement on many things, we can agree on a few things, and that is a start. We do know that there is a collection of entities in which a part or several parts of the biliary tree are abnormal in size or shape. We do know that some of these conditions can be associated with other problems, at least in some people. We do know that some people with some of these conditions are at increased risk for cancer. We know that collectively, these entities are more common in women than in men and that many, if not all, of these have a congenital component to their development.[1,2]

The fact that we cannot agree on how to classify these entities or agree on their significance or even agree whether or not they are cysts should not demoralize us. On the contrary, it should inspire us and make us curious. It matters not whether we call these entities biliary cysts or congenital choledochal malformations. It does not matter if we

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include every historical example into 1 classification scheme or another. All that matters is that we carefully assess what we do know and, perhaps more importantly, do not know about these clinical problems.

The purpose of this article is to review the information that is available and analyze through the prism of what else we have learned. At the conclusion of this article, I suggest an alternative scheme to the most widely accepted classifications. The purpose of this alternative scheme is to better match the anatomic findings with the required treatment based on what we have learned to date.

CLASSIFICATION SCHEMES

Under most circumstances, when one is writing an article such as this, one would begin with a description of the anatomy and physiology of the organs in question. It is a time-honored tradition and usually makes sense. In the case of biliary cysts or congenital choledochal malformations it is probably not so useful, perhaps even counterproductive. By definition, all of the cystic abnormalities either are or derive from anatomic variations. Furthermore, some of the defects may be the direct result of a physiologic abnormality, either de novo or as a result of anatomic variation. So, in the case of biliary cystic conditions, it makes more sense to review the standard schemes first (even if they are probably unhelpful) to understand how we got to where we are in our understanding of these entities. Once we have a grasp of the classification schemes, we can break down the entities into agreed subtypes to be followed by a, it is hoped, more useful grouping based on clinical significance.

Biliary cysts have been recognized for some time. Before the development of computed tomography (CT), fiber-optic flexible endoscopy, transcorporeal ultrasonography (US), endoscopic US, and magnetic resonance imaging (MRI), these abnormalities were identified at the time of operation, sometimes with a preoperative suspicion of their presence, other times not. The main evaluative tools were the surgeon's wits and intraoperative cholangiography. All of these diagnostic tools are readily available in many centers, as well as some variations on the themes. It is rare to stumble into a situation in which a biliary cyst is present if one is careful in one's preoperative evaluation.

The most common classification scheme currently used is the 1977 Todani modification of the 1959 Alonso-Lej classification. Alonso-Lej's original classification provided for 4 types of biliary cysts,[3] and Todani added the fifth category (**Figs. 1** and **2**).[4] In this classification, type I cysts are the extrahepatic cystic dilatations of the common duct (**Fig. 3**). They can be fusiform or spherical and can extend from the confluence of the biliary radicals to the pancreaticobiliary junction. Type II cysts are the biliary diverticula. (I am not convinced these even exist, or if they do exist that they are cysts at all. More on that to follow.) Type III lesions are the choledochoceles (**Fig. 4**).[5] The choledochoceles are frequently and erroneously referred to as type III choledochoceles. That is just wrong. One can either refer to them as a type III choledochal (or biliary) cyst or a choledochocele. There is a different, further subclassification of choledochoceles that is explored later, but it is not part of Alonso-Lej's or Todani's schemes. Type III biliary cysts are completely located within the duodenal wall and may have separate or combined entrances of the distal bile duct and ventral portion of the pancreatic duct (PD). Type IV choledochal cysts are present as multiple cysts, and at least 1 of them involves the extrahepatic bile duct. If more than 1 cystic area exists, the classification is used and further divided into type IVa and type IVb. Type IVa biliary cyst refers to cysts of the extrahepatic bile duct seen in conjunction with at least 1 intrahepatic biliary cyst. Type IVb biliary cyst refers to multiple

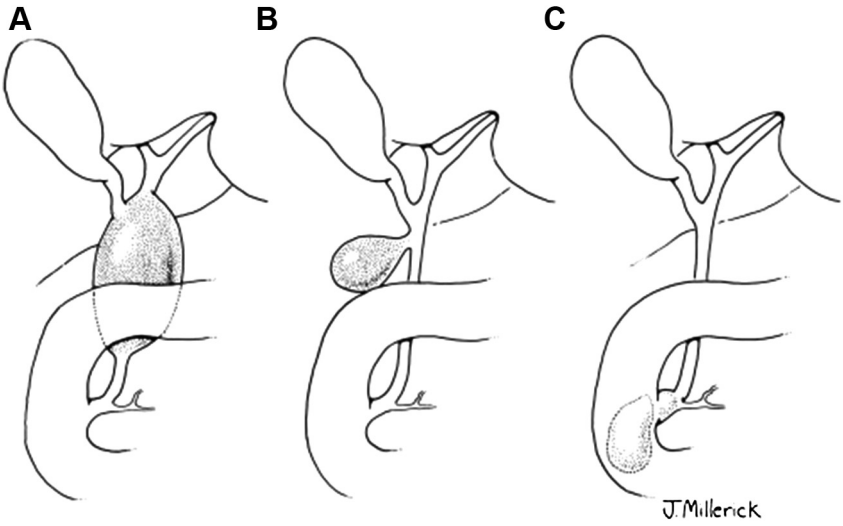

Fig. 1. Types I (*A*), II (*B*), and III (*C*) biliary cysts. (*From* Deziel DJ, Rossi RL, Munson JL, et al. Cystic disease of the bile ducts: surgical management and reoperation. Probl Gen Surg 1985;22(4):468; with permission.)

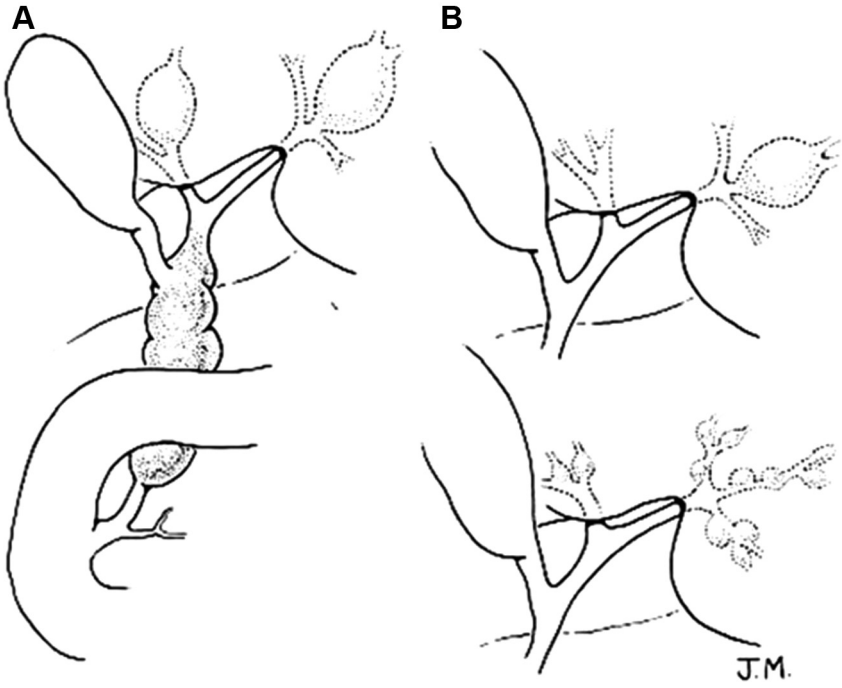

Fig. 2. Type IV (*A*) and V (*B*) biliary cysts. (*From* Deziel DJ, Rossi RL, Munson JL, et al. Cystic disease of the bile ducts: surgical management and reoperation. Problem Gen Surg 1985;22(4):469; with permission.)

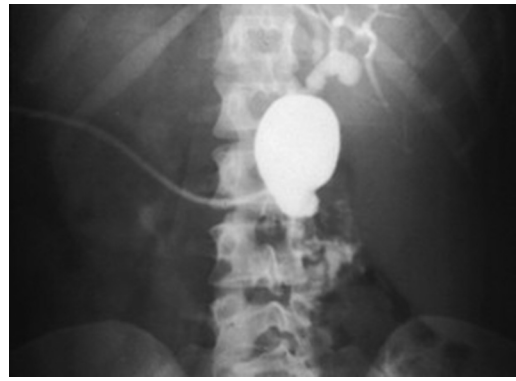

Fig. 3. Cholangiogram of type I biliary cyst. (*From* Mesleh M, Deziel DJ. Bile duct cysts. Surg Clin North Am 2008;88(6):1373; with permission.)

extrahepatic biliary cysts with no evidence of intrahepatic biliary cyst. This category would have concluded Alonso-Lej's classification scheme. Todani's modification added in type V biliary cysts. Type V biliary cyst(s) can include 1 or more intrahepatic biliary cysts but are not associated with extrahepatic biliary cysts. They may be distributed in a portion of the liver or be distributed throughout the liver. Type V biliary cyst is also known as Caroli disease (Fig. 5).

CAUSE

There are a few theories about the cause of choledochal cysts. Agreement on them is far from unanimous. The fact that the anatomic and physiologic aberrations exist

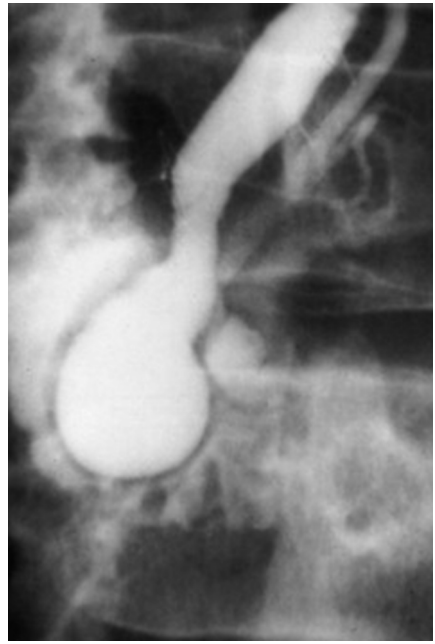

Fig. 4. Cholangiogram of type III biliary cyst. (*From* Mesleh M, Deziel DJ. Bile duct cysts. Surg Clin North Am 2008;88(6):1374; with permission.)

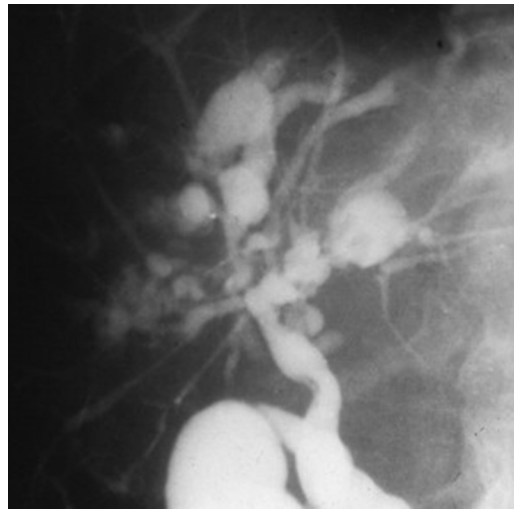

Fig. 5. Cholangiogram of type V biliary cysts. (*From* Mesleh M, Deziel DJ. Bile duct cysts. Surg Clin North Am 2008;88(6):1374; with permission.)

more so in some of the subtypes than in others leads to the increased confusion on the topic.

The main theme in most of the theories is one of an altered pressure-compliance situation. Essentially, something happens that makes the biliary structure more distensible, therefore more compliant, and this may or may not be associated with abnormally high biliary pressures within that structure. Increased compliance, as manifested by weakening of the muscular portion of the bile duct wall, could also lead to dilation in the setting of normal biliary pressure. The most often cited theory is one that Babbit[6] proposed in 1969 (**Fig. 6**). He reported a series of patients with anomalous junction of the pancreaticobiliary ducts, sometimes also referred to as anomalous pancreaticobiliary junction (APBJ), who were found to have associated type I choledochal cysts. In Babbit's model, the distal common bile duct (CBD) inserts into the PD in a more or less T-shaped arrangement, which places the pancreaticobiliary junction proximal to the main sphincter mechanisms of the ampulla of Vater and the main PD sphincter (perhaps all or in part) (**Fig. 7**).[5] The resultant anatomic variation produces a situation in which pancreatic juice can freely reflux into the bile duct, because pancreatic secretory pressure exceeds hepatic biliary secretory pressure. The theory further goes on to suppose that this reflux of pancreatic juice causes mechanical distention of the bile duct as well as inflammatory changes accompanied by degradation of the mucosa and even muscular wall that make the bile duct more distensible. Others have suggested that protein plugs that form in pancreatic juice may be a contributing factor to obstruction of outflow.[7,8] Evidence that supports these theories is increased pancreatic enzyme concentration in the cyst contents, measured pressure gradients between the PD and biliary cyst, and loss of mucosa and inflammation.[9,10] The prevalence of APBJ is reported to be 60% to 90% in patients with biliary cysts. The main weaknesses of these theories are, of course, that many patients who have biliary cysts do not have the proposed anatomic derangement. Also, the chemical studies may show association but not necessarily causation. Also, there are virtually no data on the denominator of people with APBJ who may not have an associated biliary cyst.

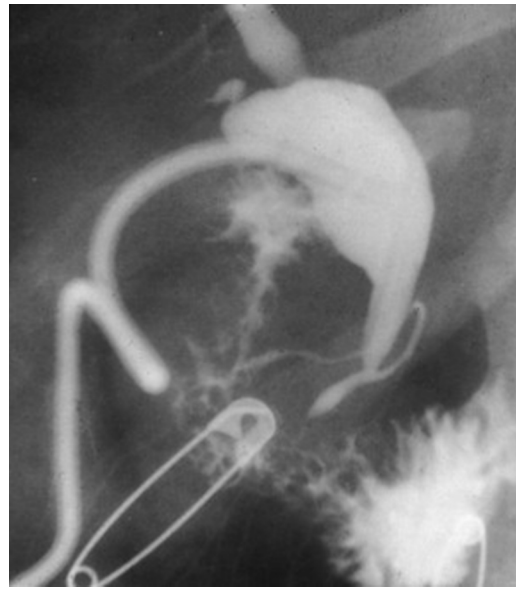

Fig. 6. Cholangiogram showing APBJ. (*From* Deziel DJ, Rossi RL, Munson JL, et al. Cystic disease of the bile ducts: surgical management and reoperation. Probl Gen Surg 1985;22(4):467–80; with permission.)

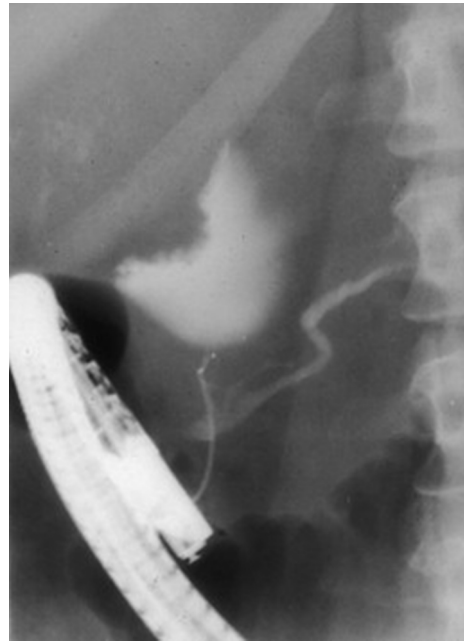

Fig. 7. Cholangiogram showing filling defect from cancer. (*From* Mesleh M, Deziel DJ. Bile duct cysts. Surg Clin North Am 2008;88(6):1379; with permission.)

Other theories of cause that do not rely completely on anomalous anatomy rely on more fundamental hydraulic principles of flow and resistance, yielding increased pressure and dilation. Animal studies have shown dilation in response to experimental ductal occlusion.[8,11,12] Others have suggested that webs or sphincter of Oddi dysfunction may be causative.[13] Embryologic variations have also been proposed. It has been postulated that the malformation of intrahepatic bile ducts in Caroli disease has been associated with faulty remodeling of the embryonic ductal plate,[14] and a maldistribution of epithelial cells within the bile duct may result in cyst formation. Motility disorders, or possible motility disorders, have also been put forth as explanatory. Tyler and colleagues[15] found that some patients with biliary cysts have fewer ganglion cells than would be expected. These investigators proposed that there might have been viral damage to the ganglion cells based on reoviral RNA levels found in patients with biliary cysts.[15] In a series of patients with type III biliary cysts, we postulated that there may be a motility disorder or motilitylike disorder associated with anomalous arrangement of the main ampullary sphincter to the distal common duct sphincter and the main PD sphincter but were unable to definitively show that.[16]

None of the etiologic theories is far fetched nor is it completely convincing. Also, none of the theories seems to tie the various types of biliary cysts (as described earlier) together. In the author's opinion, these observations make a better case for looking at the types of biliary cysts in a different way than has traditionally been described.

RISK OF MALIGNANCY

Irwin and Morrison[17] first reported the occurrence of malignancy and stones within a biliary cyst in 1944. Flanigan in 1977[18] reported a series of patients with biliary carcinomas, in whom slightly more than half had malignancy within the cyst itself (**Fig. 8**), and the others were distributed throughout the biliary system.[18] This finding led many to believe that there is a field defect associated with the presence of biliary cysts. I recommend that this assertion be taken with a large pinch of salt, because most of the cancers seen within cysts or associated with concurrent or antecedent cysts are seen in the setting of the type I or type IV cysts (ie, those with involvement of the extrahepatic duct and more likely to be associated with APBJ). Cancers are also seen in some patients with Caroli disease. The reported incidence of cancer in patients with Caroli disease is 7%.[19] Unlike the situation in type I and IV cysts with ABPJ and pancreatic reflux, patients with Caroli disease are believed to have

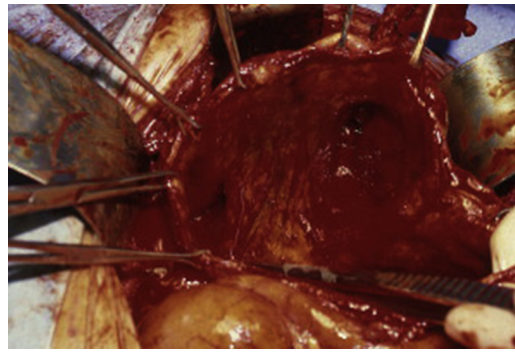

Fig. 8. Open type I cyst. (*From* Mesleh M, Deziel DJ. Bile duct cysts. Surg Clin North Am 2008;88(6):1380; with permission.)

increased risk of cancer from biliary stasis, chronic inflammation, and prolonged exposure of the bile duct epithelium to high concentrations of unconjugated, secondary bile acids.[20]

The remaining cysts are the type II and type III biliary cysts. Type II biliary cysts, or biliary diverticula, are extremely rare, bordering on nonexistent. There really is no good information on them in general and even less in regard to their association with malignancy. Type III choledochal cysts, or choledochoceles, are probably the most misunderstood of the lot of biliary cysts, which leads to confusion in any assessment of malignancy risk. The risk of cancer associated with type III cysts seems to be low. In a review by Levy and Rohrmann in 2003,[2] only 4 known cases of malignancy associated with choledochoceles were reported. A more recent review by Ziegler and Zyromski in 2011[21] found fewer than 10 such reported. In their series of 146 patients of all types of biliary cysts, these investigators reported 6 patients with associated neoplasm, but none in the group of type III cysts.

TYPE I CYSTS

Type I biliary cysts, or extrahepatic biliary cysts, are uncommon but not unheard of in the general population. They are the most common of the biliary cysts. Their incidence is estimated at 1:100,000 to 1:150,000 in Western countries and higher in Japan, at 1:1,000.[22] The knowledge of their presence dates back to Vater in 1723.[23] Douglas in 1852[24] suggested the congenital nature of the cyst. The clinical presentation of patients with type I biliary cysts may include abdominal pain, jaundice, an abdominal mass, or some combination of these. In children, pain may be the only presenting symptom. Given modern imaging techniques (and perhaps declining physical examination skills of physicians and increasing levels of obesity), palpable abnormalities preceding image identification are fairly uncommon.

As mentioned earlier, altered anatomy in conjunction with reflux of pancreatic juice or perhaps other obstructions to biliary outflow is believed to lead to the development of cystic dilatation. The type I cysts are the most common of all the biliary cysts. They are 3 times more common in females than in males[25] and may present at any age. Although mainly presenting with jaundice or pain, they may present in association with gallstones in any part of the biliary tract, cholangitis, biliary cirrhosis, portal hypertension, or malignancy.[26] The risk of associated malignancy seems to increase with age, with reported incidence ranging from 2.5% to 28%.[27–29] Todani and colleagues[29] found in their series that about half (49.8%) of the cancers that they saw were in the cyst itself and about half (46.5%) were found within the gallbladder, with the remaining 3.7% equally distributed within the liver or pancreas.

The diagnosis of biliary cyst can be made clinically, or at least suspected, but is usually confirmed by some imaging technique. Transcorporeal US is probably the most common initial imaging modality used, but with the almost ubiquitous use of CT, that may no longer be true. Almost any imaging modality raises the flag of suspicion, and many can be used to identify what is important. Body imaging such as CT, MRI, and US are all potentially useful and may define the extent of ductal involvement and presence or absence, of remote disease. If further delineation of the ductal system is required to plan treatment, endoscopic retrograde cholangiopancreatography (ERCP), magnetic resonance cholangiopancreatography, or percutaneous transhepatic cholangiography can all be used. I have not found radionuclide studies to be of major help in planning treatment or diagnosis.

The treatment of type I cysts is removal of the cyst and restoration of biliary continuity. Exceptions to this principle include patients with suspected malignancy outside

the cyst or patients who are considered unacceptable operative candidates for other reasons. Reconstruction almost always requires hepaticojejunostomy, although rarely, a reconstruction above the biliary confluence is required. This operation can be tedious and does require excellent attention to detail and operative planes. However, the operation can be performed safely, with little to no mortality and low complication rates.[30–34] The fundamental rules of biliary surgery are no different than for other operations: at the conclusion of the case, all liver segments need to have unobstructed flow of bile into the lumen of the gastrointestinal tract. This requirement implies that all steps are taken to ensure that variant anatomy is not overlooked, and all debris is removed from the remaining ducts during the conduct of the operation. The distal limit of resection also needs careful attention. Most commonly, one can inspect through the open or divided cyst and see the narrowing of the intrapancreatic portion of the bile duct. Mesleh and Deziel[5,31] emphasize oversewing the distal duct to prevent possible pancreatic fistula and also advocate the placing of drains, both suggestions with which I wholeheartedly concur. In situations in which one is concerned about maintaining the integrity of the pancreaticobiliary junction, it may be advisable to place a nasopancreatic tube before the operation. It can offer several benefits, such as being able to see or feel the tube in addition to being able to inject the tube with either radiographic contrast or optically detectable fluid during the operation if necessary. Lily[35] has suggested limited resection of the cyst to spare the portion adherent to the portal vein. Personally, I have never encountered a situation in which this would have been helpful or even easier. Careful dissection has invariably produced a safe plan between the bile duct and the portal vein that has allowed for complete excision of the cyst.

Attempts at cyst drainage procedures should be of historical interest only. The only good reason I can think of to address them in this article is that the reader may encounter a patient who has had one already. Anastomoses of the cyst to the stomach, duodenum, or jejunum without excision have all been performed, and all fail to address the main concerns of biliary cysts. The problem of pancreatic juice reflux is left unaltered completely. These procedures tend to be complicated by stricture, stone formation, and recurrent cholangitis. When one encounters a patient who has undergone a previous limited drainage procedure, the approach is the same as if it were a de novo presentation. The major differences are in technical issues regarding evaluation and treatment based on how the anatomy has already been altered.

TYPE II CYSTS

Type II cysts, or biliary diverticula, are rare. They are so rare that, in my opinion, it would be best to discontinue including them in the discussion of biliary cysts. The differential diagnosis includes duplication of the gallbladder. Intersphincteric diverticula may represent a herniation of bile duct mucosa and, therefore, be false diverticula akin to sigmoid diverticula, whereas the more proximal diverticula are believed to be congenital true diverticula. The only published cholangiograms showing what one could construe as a biliary diverticulum that I have ever found are contained in a report from the Armed Forces Institute of Pathology.[2]

The treatment of these lesions needs to be based on fundamental principles as opposed to any real data. We have no idea if they represent a malignant risk or, if left alone, anything of adverse consequence would occur. Therefore, the main reason to intervene would be if someone were to present with symptoms or signs of clinical problems attributable to the diverticula. In that case, excision with either maintenance of biliary drainage or restoration of biliary continuity would be in order. In my opinion, if

we all collectively decided to exclude this entity from discussion, the net impact would be negligible.

TYPE III CYSTS

Type III cysts, or choledochoceles, are located within the duodenal wall. The term choledochocele was coined by Wheeler in 1940.[36] It is believed to be the second least common of the biliary cysts, although I think that we should reserve judgment on that. The differential diagnosis includes duodenal duplications and other cystic abnormalities of adjacent anatomic structures. Further confusing the topic is that some investigators include cystic abnormalities with duodenal mucosa[37] as well as biliary mucosa.[38,39] The cysts range in size from small to large enough to palpate on examination. The very large ones can obstruct the duodenum, whereas others may present with jaundice or persistent or recurrent acute pancreatitis.

Scholz and colleagues[37] described a scheme for choledochoceles that differentiates based on separate insertions of the CBD and PD or a common channel of CBD and PD inserting into the cyst: types A and B, respectively. Sarris and Tsang[40] proposed a more complicated scheme of choledochoceles, which combines common and noncommon channels as well as intramural versus intraluminal characteristics for classification. In our series, we found that all of the choledochoceles that we encountered had biliary mucosa and a separate entrance of the CBD and PD.[16] We also discovered a radiographic finding of parallel folds within the cysts, which we referred to as Biber lines. We postulated that there was a defect in the crossing fibers of the sling mechanisms between the sphincter of Oddi and the main CBD and main PD sphincters that allows for bulging between the sphincters, allowing for a bulging cyst to develop. We were not able to prove this theory, because we did not have manometric data or deep wall biopsies to show altered ganglion presence or dropout of the muscular sling.

It seems logical that the duodenal mucosa–lined cysts (duplications) and biliary mucosa–lined cysts (probably what we think of as a choledochocele) should behave differently. As stated earlier, the relationship, if any, to neoplasia or proper cancer is entirely unclear, and it is at most rare. Lesions that are large enough to cause obstruction of the duodenum may need to be managed by operative means, usually with transduodenal excision, which may or may not require reimplantation of the CBD and PD. Smaller lesions may be managed via unroofing only, via operative means or via endoscopic means, as was performed in our series.[16]

I have previously suggested that we may not know the true incidence of choledochoceles. The reason I suggest this is, since the advent of readily available flexible fiber-optic endoscopy and ERCP, it is entirely likely that many bulging papillas that have been managed with endoscopic retrograde sphincterotomy or needle-knife papillotomy may have been early or small choledochoceles. Even if so, the distinction would be largely academic and of no true clinical significance.

TYPE IV CYSTS

Type IV biliary cysts are classified separately, because multiple cysts are present, at least one of which must be extrahepatic. If the multiple cysts are extrahepatic and intrahepatic, they are considered type IVa cysts, and if the multiple cysts are all extrahepatic, they are considered type IVb cysts. This is probably an unnecessary classification, because it adds little to help us. In the category of the type IVb cysts, everything that can be said about type I cysts also applies without exception. In the category of the type IVa cysts, what is said about type I cysts applies as well to the

extrahepatic component, and what is stated about type V biliary cysts (Caroli disease) applies to the intrahepatic portion.

I refer the reader to the sections on type I and type V cysts, because repeating the information here is counterproductive. This is 1 more reason why I add my support to the abandonment of this classification scheme later in this article.

TYPE V CYSTS

Caroli disease is also known as communicating ectasia of the intrahepatic bile ducts. It is a rare autosomal-recessive disorder described by Caroli and others in 1958.[41] As mentioned earlier, it is a result of embryologic ductal plate malformations that occur during remodeling. The resultant defect can cause fibrosing and scarring of bile ducts large or small. When the scarring is limited to the smaller duct, the effect may be negligible, but when the scarring involves the larger ducts, the large cystic collections that define Caroli disease can form.

Renal cysts are seen with disorders with the ductal plate, and renal developmental abnormalities can be caused by the same genetic determinants.[20] Autosomal-dominant and autosomal-recessive polycystic kidney, as well as medullary sponge and medullary cystic kidney, can be seen in association with Caroli disease.[2]

The treatment of Caroli disease is based on removing the portion of the liver containing the cysts and any residual impediment to biliary drainage of remaining segments. The treatment needs to be tailored to the patient being considered.[14,42] Factors that need to be taken into account are the extent and distribution of the cysts and associated strictures, any associated malignancy, as well as the underlying function of what will be the residual liver. Of course, the general medial condition of the patient may be determinant in the patient's ability to withstand a significant operation. Transplantation is the fallback position when there is either pan-lobar involvement or predicted inadequate functional residual liver after partial liver resection. If the distribution of disease is such that it can be eliminated with segmental resection or multiple segmental resections, yielding adequate residual liver mass, then this should be preferable to transplantation. There are insufficient data to suggest that any given operative approach to these resections is better than another, whether performed by conventional operative means or videoscopic or robotic means.

A SIMPLIFIED APPROACH

I submit to you, dear reader, that from this discussion, we have enough information to generate a simpler approach to biliary cystic disease than is currently used. In no way do I wish to denigrate the work of all those who have toiled before us. After all, it was their work that allowed us to study these disorders and learn what we have. Now, it is time to make use of that effort. I am not the first nor will I likely be the last to suggest an alternative classification scheme, but I provide one for us to consider.

The basis of the change is simple. Based on what we know to date, the type I to V classification scheme is overly complicated and occasionally confusing. If we begin by eliminating the biliary diverticula altogether, because they are exceedingly rare to the point of near nonexistence, that leaves us with 4 residual classes of cysts. (For anyone who fears or laments the loss of the type II cysts, we can give them a new place in surgical discourse away from here, much the way that Pluto lost its planetary status; Pluto, after all, is still there; it is just not called a planet anymore.) The type IV class of cysts is at best redundant. The type IVb are really just odd type I cysts with all the same clinical consequences and same treatment options. The type IVa cysts

Table 1
Simplified classification scheme for biliary cysts

Type	Treatment
Intrahepatic cysts	Segmental resection where possible and the patient is an adequate operative candidate; consider transplantation if inadequate liver reserve is expected
Extrahepatic bile duct cysts	Resection of extrahepatic bile ducts (including complete resection of cyst(s)) with restoration biliary continuity
Intraduodenal cysts	If small cyst unroofing by endoscopic or operative means if possible; if larger or unroofing not possible, resect cyst via transduodenal approach, possibly requiring reimplantation of CBD or PD

are just rare combinations of type I and type V cysts, which may be related abnormalities or coincidental abnormalities. Whatever advice is used for type I or type V cysts can be used to guide one when treating patients with these cysts. That leaves us with intrahepatic cysts and extrahepatic cysts. Honestly, that is it. The last point we need to address is whether or not to include the choledochoceles or type III cysts. Ziegler and Zyromski[21] have made a compelling argument for the exclusion of choledochoceles from the list of biliary cysts. Perhaps because of my previous work, or just not knowing where else to orphan these entities (because they are more prevalent than type II cysts), I support keeping them in for now.

If one keeps to this system, one is left with a scheme that includes intrahepatic cysts, extrahepatic cysts, and intraduodenal cysts. The treatment paradigm that would follow is shown in **Table 1**. For intrahepatic lesions, resect as little liver as possible to eliminate the cysts and accompanying strictures; however, as little as possible may require complete liver resection and transplantation. For extrahepatic lesions, resect the bile duct(s) that are involved and restore biliary drainage. For patients with both intrahepatic and extrahepatic cysts, follow each of these algorithms. For patients with intraduodenal lesions, unroof those smaller lesions by either endoscopic or operative means if possible. For larger lesions, resection via transduodenal approach with possible reimplantation of CBD or PD may be required.

SUMMARY

There is much known, and there is much unknown, about biliary cystic disease. Several classification schemes have been developed and modified over the last 70 years or so. What seems to be true is that the various biliary cystic conditions that have been considered together represent markedly different disorders in terms of embryologic and anatomic considerations, and they also carry different risks of both neoplastic and nonneoplastic complications. Modern imaging techniques, as well as advances in operative technique and our ability to manage patients in the perioperative period, have likely altered and simplified the necessary classification schemes and treatment algorithms.

As I stated at the beginning of this article, if you know nothing about biliary cysts, study a little, and you will know a lot. If you study a lot, you will soon realize how little we really do know, probably a metaphor for life. I have tried to demystify this situation as best as possible. Time will tell if I have oversimplified this or have completely missed the point. I look forward to the day that one of you revises this article and tells us how far off the mark I was.

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