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