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Management of extrahepatic bile duct cysts

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Introduction

Bile duct cysts are rare anomalies that to this day are poorly understood. They are more common in the Asian population, and often found and treated in childhood[1]. In Western countries, bile duct cysts are more commonly discovered incidentally in adults or as part of work-up for jaundice or non-specific upper abdominal symptoms[2–4]. Often referred to as 'choledochal cysts', even in contemporary literature, the biliary dilatation can occur throughout the intrahepatic and extrahepatic biliary tree. Thus, the term 'bile duct cyst' or 'biliary tract cyst' is preferred.

How to classify bile duct cysts

Several classifications have been proposed for bile duct cysts, the most recent being that of Todani *et al.*[5] (*Fig. 1a*). Type I cysts are limited to the extrahepatic ducts and make up 70–90 per cent of all cysts. The second most common is type IV, accounting for 10–15 per cent, comprising multifocal cysts, either with an intrahepatic component (type IVA) or only extrahepatic (type IVB). Type II cysts present as a diverticulum at the common bile duct, and type III is a cyst in the intrapancreatic part of the distal bile duct. Some would argue that types II and III should not be classified with the other cysts as true congenital cysts[6].

Type V cysts (also called Caroli's disease) are intrahepatic and multifocal. Of note, recognizing a dilated biliary duct as a true cyst may be difficult[7].

What is a pancreatobiliary maljunction?

A pancreatobiliary maljunction, also referred to as long common channel or anomalous pancreatobiliary ductal union, is a congenital malformation in which the bile and pancreatic ducts join outside the duodenal wall (*Fig. 1b*)[8–10]. The theory is that the maljunction facilitates pancreatic enzyme reflux into the biliary tree causing persistent inflammation, which could explain both the creation of cysts and the increased risk of malignancy over time[8,11]. Seventy per cent of bile duct cysts are associated with the presence of a pancreatobiliary maljunction[12]. Of note, there is an inconsistent presence of the two designated anomalies, as each can be found without the other. The causality between cancer risk and the presence of a pancreatobiliary maljunction is unclear, as the presence of such an anatomical anomaly could represent a mere anatomical association[8,9,13–16]. In one comparative study[13] of patients from North America and Japan who underwent resection for hepatobiliary and pancreatic malignancy, about 8 per cent had a pancreatobiliary maljunction, suggesting the prevalence to be comparable between East and West for this anomaly[12]. In addition, a higher risk of associated inflammatory conditions, such as cholangitis, cholecystitis, and pancreatitis, as well as stone formation, seems to be a consistent effect of having a pancreatobiliary maljunction.

What are the risks of having bile duct cysts and when should they be resected?

Bile duct cysts are associated with an increased risk of developing biliary tract cancers[11,17,18], reported in about 10 per cent. The risk increases with age, and such cancers often present a decade earlier than spontaneous cholangiocarcinoma. The highest risk of malignancy is in the most prevalent types I and IV. Of note, type II and type III cysts (less than 1–2 per cent of all cysts) may not be associated with reflux of pancreatic secretions into the bile duct, and as such have a much lower or even non-existent carcinogenic risk. The risk of cholangiocarcinoma in Caroli's disease was 6–7 per cent in a German multicentre study[19].

How to best diagnose and classify biliary tract cysts

MRI with magnetic resonance cholangiopancreatography (MRCP) is the reference standard to best diagnose and classify the cysts (*Fig. 2*), but also to designate any concomitant pancreatobiliary maljunction[20–22]. Changes in diameter, appearance of contrast-enhanced intracystic nodules, and restricted diffusion in the setting of a cyst with a pancreatobiliary maljunction may suggest risk of malignancy. Routine endoscopic investigation with endoscopic retrograde cholangiopancreatography (ERCP) is usually not needed if appropriate MRCP is done, unless there is suspicion of obstruction or cholangitis[12,22]. ERCP may, however, help to facilitate the diagnosis in some cases of diagnostic uncertainty, or to achieve cyst fluid aspiration, brush cytology, or

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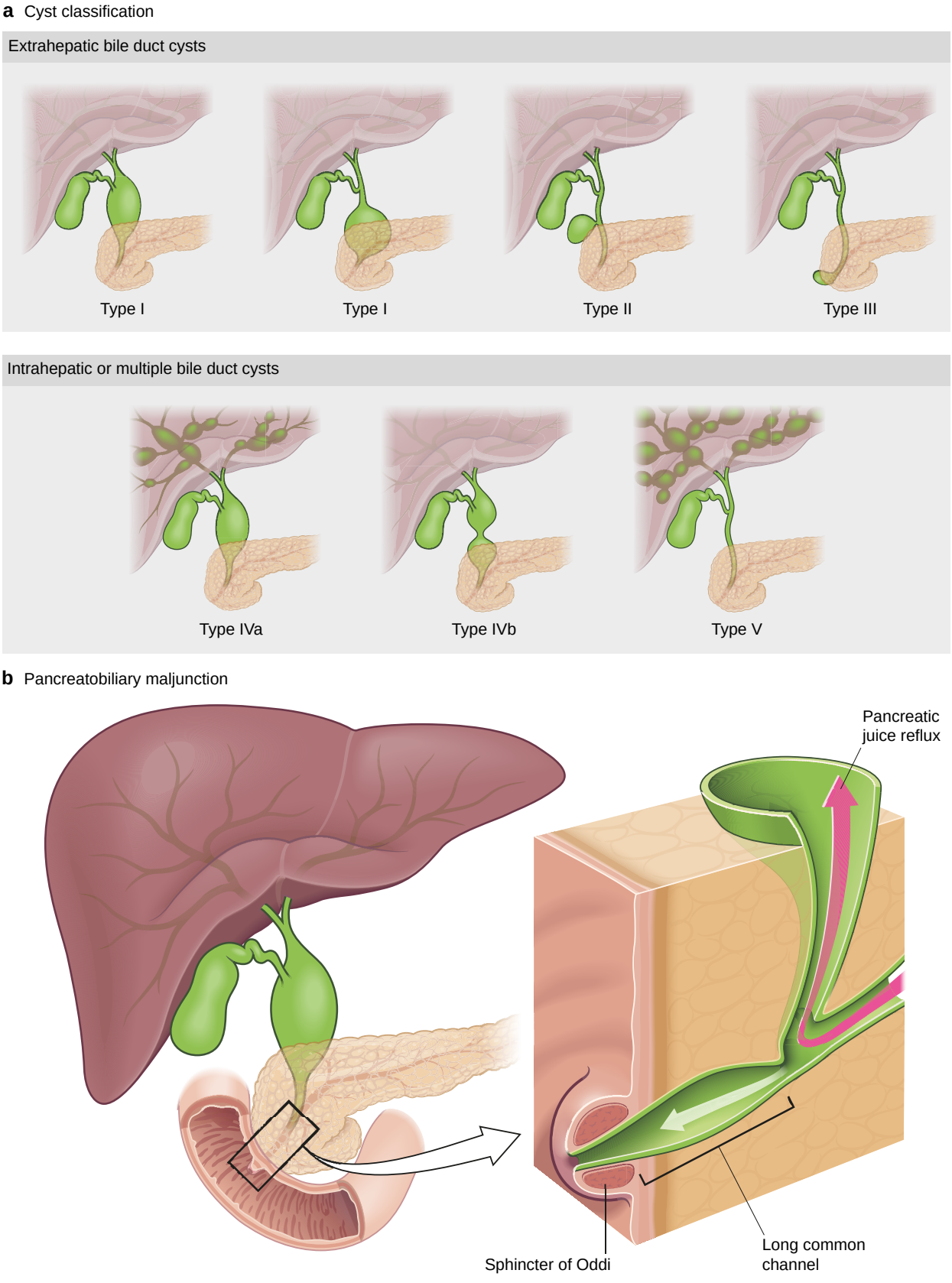

Fig. 1 Classification of bile duct cysts and depiction of a pancreatobiliary maljunction

a Types of cyst classed as I, II, III, IV and V. Type I is a fusiform or saccular type; type II is a diverticulum; type III (choledochocele) is located in the duodenal part. Type IV cysts may involve intrahepatic parts of biliary tree (type IVA) or may be multifocal, extrahepatic only (type IVB). Type V (Caroli's disease) denotes bilobar intrahepatic cystic dilatations. **b** A maljunction is present either when there exists a specific angle at which the bile duct joins the pancreatic duct that predisposes to reflux of pancreatic juices into the bile duct, or when there is a long common channel (usually defined as over 8–10 mm) between the junction of the ducts and the sphincter of Oddi, creating a pressure that propagates reflux retrogradely into the bile duct.

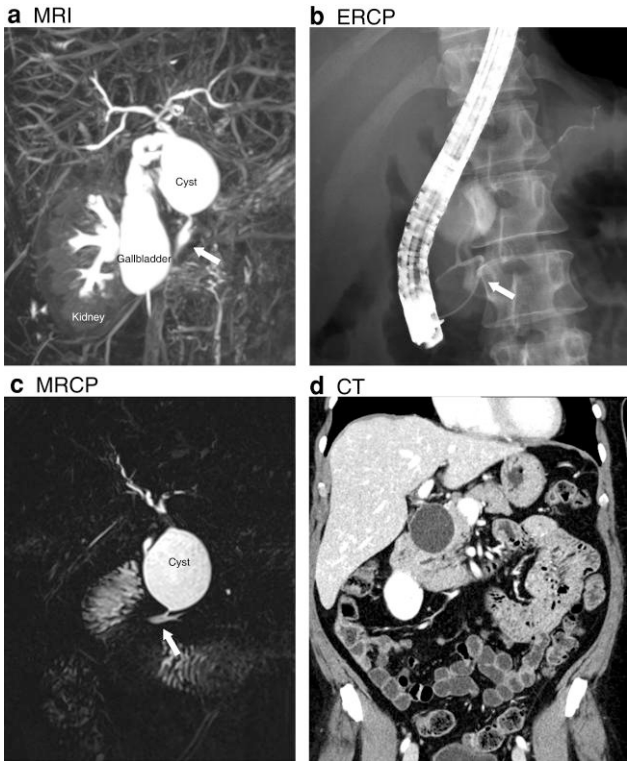

Fig. 2 Imaging for bile duct cysts

a MRI of Todani type I cyst, with long common channel (arrow). **b** Endoscopic retrograde cholangiopancreatography (ERCP) showing type I cysts with a long common channel (arrow). **c** Magnetic resonance cholangiopancreatography (MRCP) showing type I cyst (saccular type) and a long common channel (arrow). **d** CT showing type I cyst (saccular type) of distal bile duct with intrapancreatic involvement.

cholangioscopically guided biopsy. Furthermore, type III cysts (choledochocele) may be treated by endoscopic sphincterotomy rather than resection[6]. Cyst fluid aspiration to measure amylase levels during ERCP or percutaneous drainage is a controversial topic[23–26]. The idea is that amylase reflux increases the risk of malignancy. Such reflux is, however, found in both people with and those without pancreatobiliary maljunction. Although the cancer risk is clearly greater with high amylase levels, it is also considerable at lower or normal ranges[23].

Which operation and reconstruction?

Drainage procedures such as cystoduodenostomies are not recommended. They have been shown to be associated with a higher risk of metachronous cancer than resectional procedures[4,17].

A type I cyst requires resection of the affected segment of the bile duct with a Roux-en-Y hepaticojejunostomy. The challenge with these resections usually is at the level of the distal resection margin, as controversy remains regarding the degree of radicality of the intrapancreatic part of a cyst. Going into the pancreatic parenchyma may cause a fistula or later stricture of the pancreatic duct, whereas leaving tissue behind may put the epithelium at risk of future cancer development. Pericystic fibrosis is not uncommon and may further complicate this evaluation.

Surgery for type II and type III cysts is controversial, and should be avoided unless symptoms of biliary obstruction or recurrent pancreatitis are documented. A type II cyst is a true diverticulum and can be resected at its neck where it joins the common bile duct, whereas type III cysts (choledochocele) can be treated endoscopically with a sphincterotomy.

Resection for type IV cysts can necessitate simultaneous liver resection when the cysts involve both the intrahepatic and extrahepatic ducts. Type V cysts (Caroli's disease) are usually treated by liver transplantation, unless there is a defined segment affected, which may then be safely resected.

Why and how to undertake long-term surveillance?

Follow-up after surgery serves two purposes: to monitor for development of malignancy in the remnant biliary tissue, and development of late complications, such as anastomotic strictures or formation of stones or debris. Metachronous cancer is reported to occur in 2–4 per cent of patients after resection[27,28]. The prevalence of non-malignant strictures is reported to range from 2 to 17 per cent, a problem that appears to be less of an issue among patients operated in childhood[29]. Follow-up may consist of annual liver function tests and carbohydrate antigen 19-9 measurement. Based on availability, the authors also recommend cross-sectional imaging with MRCP.

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Disclosure

The authors declare no conflict of interest.

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