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REVIEW ARTICLE

UPDATES IN THE MANAGEMENT OF CHOLEDOCHALCYSTS IN ADULTS

By

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Choledochal cysts are congenital anomalies of the bile ducts. They consist of cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary radicles, or both. They can occur as single or multiple cysts. Choledochal cysts account for less than 1% of all benign biliary diseases ***(Singhavejsakul and Ukarapol, 2008)***.

The incidence of choledochal cysts is about 1:100,000-150,000 in western countries with male to female ratio of 1:3. It is more frequently seen in Asian countries, especially Japan, where the incidence is about 1:1,000 with a male to female ratio of 1:8 because of the effect of the sex steroid on the sphincter of oddi and the lithogenic consistency of the biliary secretion ***(*Aguilera et al.*, *2004)***.

The pathogenesis of choledochal cysts is most likely multifactorial. Some aspects of the disease are consistent with a congenital etiology, and others with a congenital predisposition to acquire the disease under the right conditions. The most widely accepted theory for development of choledochal cyst is anomalous pancreaticobiliary duct junction ***(Nicholl et al., 2004)***.

Todani et al. (1977) modified the classification system of bile duct cysts by combining the Alonso-Lej classification system and variants of Caroli's disease. The main criteria based on are cholangiographic morphology, location, and number of intrahepatic and extrahepatic bile duct cysts (Fig.1).

The classic type and most common choledochal cyst is the type I choledochal cyst. It accounts for 50-80% of choledochal cyst patients. It is characterized by cystic or diffuse fusiform dilation of the entire common hepatic and common bile ducts, or of segments of each, with the fusiform type being more common. Type II choledochal cysts are relatively isolated protrusions or diverticulae of the extrahepatic biliary tree located proximal to the duo denum and are extremely rare, accounting for fewer than 5% of all choledochal cysts. A choledochocele, or type III choledochal cyst, is a cystic dilation of the intraduodenal portion of the biliary tree. It accounts for approximately 5% of choledochal cyst patients. Type IV choledochal cysts involve multiple dilations of the intrahepatic and extrahepatic biliary tree. Type IV biliary cysts are further subdivided into type IV_A

(multiple intrahepatic and extrahepatic cysts), and IV_B (multiple extrahepatic cysts without intrahepatic involvement). Type IV_A is the second most common type of biliary cyst, and accounts for 30-40% of choledochal cyst patients. Caroli's disease, or type V choledochal cyst, is confined to the intrahepatic portion of the biliary tree. It accounts for less than 1% of choledochal cyst patients. ***(Todani et al., 2003)*.**

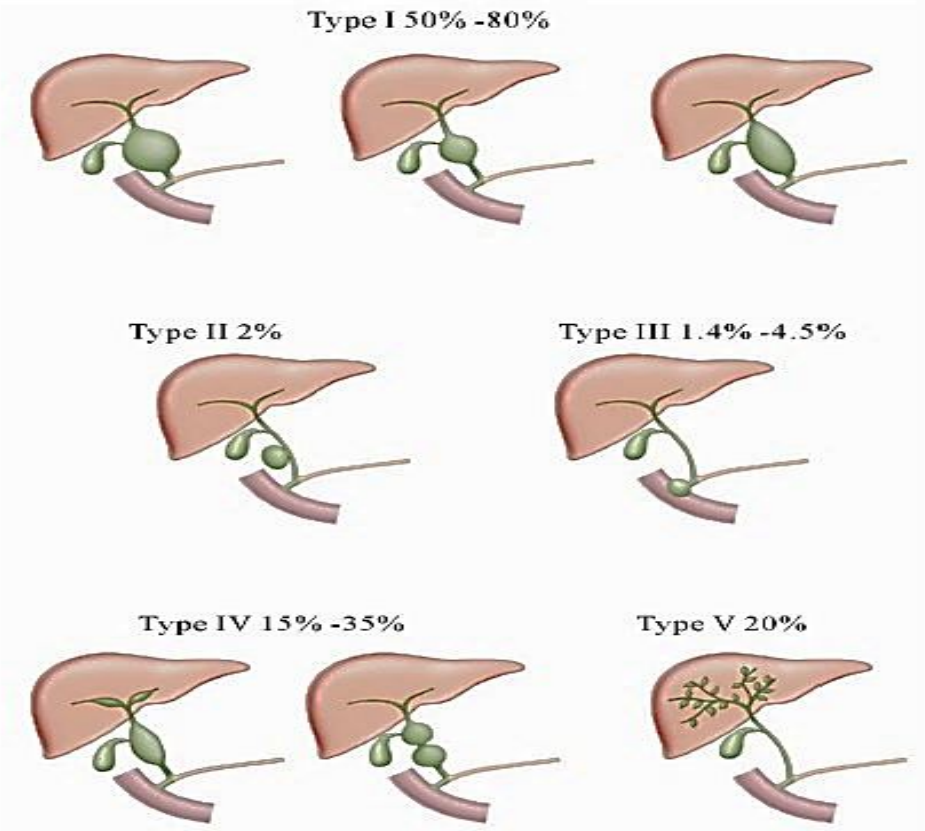

Figure (1): Todani classificationof choledochalcysts***(Woon et al., 2006)***.

Choledochal cyst can manifest at any age, from gestation to old age. Typically, it presents during infancy and childhood, and more than 60% of all cases are diagnosed in the first decade. Unfortunately, the diagnosis is delayed in approximately 20% to 30% of cases, often with adverse consequences. Choledochal cyst presents with different clinical symptoms determined by patient age, cyst type, and presence or absence of associated hepatobiliary complications. ***(Leeet al., 2005)***.

Jaundice is the main presenting symptom of extrahepatic cysts, cholangitis, and gallstones of intrahepatic cysts. This may be explained by the localization of the lesion. Extrahepatic cysts may give complete obstruction of the biliary tree leading to jaundice, whereas intrahepatic cysts will lead to partial obstruction giving late and

localized complications ***(Todani et al.,2003)***. It is found that cholangitis is the most common manifestation of Caroli's disease (about 50% of patients).

When symptomatic, choledochal cysts usually present similar to calculus biliary tract disease, regardless of cyst type. Symptoms typically are intermittent, as most of these patients have fusiform deformities of the common bile duct with partial obstruction.

The formation of mucus plugs and biliary sludge are associated with appearance of symptoms. Recurrent epigastric or right hypochondrial pain, abdominal tenderness, fever, and mild obstructive jaundice are the most common presenting findings. The pain may radiate to the right infrascapular region or to the mid back, and generally persists for hours.Abdominal pain or discomfort is often overshadowed by fever and rigors, which may occur repeatedly for several days.

Choledochal cysts are associated with many different developmental anomalies, which have given to rise some additional etiological theories. Such associations include colonic atresia, duodenal atresia, imperforate anus,familialadenomatous polyposis, pancreatic arteriovenous malformation, multiseptate gallbladder, biliary atresia, ventricular septal defect, aortic hypoplasia, pancreatic divisum, pancreatic aplasia, and focal nodular hyperplasia.

However, the presence of various cysts related complications may alter the classical presentation and also influence the subsequent management and outcome.

Complications related to choledochal cysts have been broadly divided into infective and non-infective complications.

Acute suppurativecholangitis, acute cholecystitis and intra hepaticabcesses are the infective complications that could occur in the cases of the choledochalcysts.

Spontaneous perforation of the cyst,cystolithiasis,hepaticolithiasis and recurrent acute pancreatitis are noninfectivecomplications.
Portalhypertension, gastric outlet obstruction and malignancy are late reported complications (***Germiller et al., 2007)***.

Diagnosis of the choledochal cysts is dependant on the clinical symptoms and possible complications listed before. Besides, laboratory and radiological investigations are initial and confirmative tools respectively in the diagnosis.

The bilirubin is usually elevated to some degree in obstructive pattern (conjucated hyperbilirubinemia), with elevated alkaline phosphatase and gamma glutamyl transferase. The transaminases (alanine and aspartate aminotransferases – ALT&AST) can be elevated to a lesser degree. If biliary obstruction has been present for a substantial period, patients may even have an abnormal coagulation profile, hypoproteinemia and hypo-albuminemia .Serum amylase may be elevated in patients presenting with acute abdominal pain and signs and symptoms of clinical pancreatitis.***(Parket al., 2005).***

Ultrasound (US) is the tool of choice for diagnosing biliary disorders. Once a preliminary diagnosis is made using US scaning, other supportive studies may be ordered, including abdominal computed

tomography (CT) scans, magnetic resonance imaging (MRI) studies, magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP), and percutaneous transhepatic cholangiography (PTC) examinations. These studies demonstrate the cyst with more precise anatomic detail, important anatomic relationships to surrounding structures, precise definition of the biliary anatomy, and associated complications. ***(Oto et al.,2009).***

In children and adults, choledochal cyst may be confused with several other cystic lesions including hepaticcysts, hepatic artery aneurysm gallbladder duplication, spontaneous perforation of the common bile duct, choledocholithiasis, cystic biliary neoplasms such as biliary cystadenoma and cystadenocarcinoma, pancreatic cysts and pseudocysts, enteric duplication cyst, and mesenteric cysts**(Woo et al., 2006).**

The definitive treatment of choledochal cyst is surgery. The surgical management of choledochal cyst is fashioned depending on multiple factors. Factors to be considered when performing surgery on patients with choledochal cyst includeage, presenting symptoms,cyst type, associated biliary stones, prior biliary surgery, intrahepatic strictures, biliary cirrhosis, portal hypertension, and presence of associated hepatobiliary pathology especially malignancy***(Lipsett et al., 2003)***.

The aim of preoperative management is complete cholangiographic definition of the extent of the cystic process and associated ductal pathology, and control of biliary infections. Patients in whom sepsis fails to resolve with intravenous antibiotics require preoperative percutaneous or endoscopic drainage of infected choledochal cyst, which usually affords control of sepsis before definitive operation. In general, all choledochal cysts should be excised, and bile flow re-established by mucosa to mucosa biliary-enteric anastomosis. If complete excision is not feasible, partial cyst excision and Roux-en-Y cystojejunostomy to an epithelial lined portion of the cyst remenant is preferred. External drainage alone has no definitive role in the surgical management of choledochal cyst. Routine cholangioscopy is employed in adults to exclude retained ductal stones and ductal malignancies· In general, regardless of age, presenting symptoms, biliary stones, prior surgery, or other secondary problems, surgery should include cholecystectomy and excision of extrahepatic cyst(s).

The theoretical requirements of an ideal operation are:

(1) To allow free hepato-enteric bile flow.
(2) To remove all cyst mucosa (with its associated malignant potential).
(3) To minimize the subsequent risk of cholangitis. ***(Shimotakahara et al.,2005)***.

Resection of the intrahepatic and intrapancreatic portions of the cysts reduces the risk of cancer even though this risk is low after incomplete cyst excision. Biliary continuity after cyst resection is best established by Roux-Y hepaticojejunostomy.

Minimally invasive surgery has evolved as a standard technique for the treatment of numerous conditions. Laparoscopy has gained enormous

popularity in the management of cholelithiasis. Yet, its application in the surgical excision of choledochalcyst was very limited. Laparoscopic choledochal cyst excision and hepaticojejunostomy for Type I and Type II cysts has been described as an alternative to open surgery. Laparoscopy may not be feasible in types IV and V, where there is an intrahepatic component **(Srimurthyand Ramesh, 2006).**

Proper case selection is crucial for a good outcome. A certain degree of caution has to be exercised before embarking on laparoscopic repair of choledochal cyst. The surgical and the anesthetic team and the supporting staff should have sufficient experience in advanced minimal invasive surgery. Difficulty may arise in older patients where the size of the cyst may be very large and adherent to surrounding structures. Biliary anatomy is distorted as a result of adhesions and cystic dilation, which makes dissection particularly difficult. The dissection may be cumbersome due to inflammation and bleeding in cases with a prior history of cholangitis,and it may be prudent to avoid such cases for laparoscopic surgery. Caution is needed in patients with associated liver cirrhosis and portal hypertension. Extreme caution has to be exercised during the dissection to avoid troublesome bleeding, which will obscure vision. The dissection of the lower end has to be very meticulous down to the lower extent of the choledochalcyst. This is possible if the lower end of the cyst is gradually extracted out of the duodenum. A sufficient margin of the proximal cuff is essential for easy anastomosis. The use of a 30 or 45 telescope aids suturing***(S?reide et al., 2004)***.

Woo (2006) experimented with a robot-assisted laparoscopic resection of a type I cyst in 8 patients. Laparoscopic resection has not been popular because of the technical difficulties with performing the hepaticojejunostomy. The robot-assisted technique did simplify the most complex part of the procedure compared with standard laparoscopy, but the cost and the robotic training requirements make it unlikely thatit will replace the open method **(Srimurthy and Ramesh, 2006).**

Long term follow up after treatment is essential because recurrent cholangitis, lithiasis, anastomotic stricture and pancreatitis may develop within years after the initial operation.

REFERENCES

1. **Aguilera V., Ray?n M and Pérez-Aguilar F. (2004):** Caroli's syndrome andimaging: report of acase. *Rev Esp Enferm Dig*., 96:74−76.
2. **GermillerJA., StrousePJ and GolladayES. (2007):** Early presentation of choledochalcyst transiently obstructed by an inspissated bile plug. *J Pediatr Surg*., 32:1522−1525.
3. **Lee KF, Lai ECH and Lai PBS. (2005):** Adul tcholedochal cyst. *Asian J Surg*., 28:29−33.
4. **Lipsett PA and Pitt HA. (2003):** Surgical treatment of choledochal cysts. *J Hepatobiliary Pancreat Surg* .,10:352−359.
5. **Nicholl M., Pitt HA and Wolf P. (2004):** Choledochal cysts in western adults: complexities compared to children. *J Gastrointest Surg*., 8:245-252 .
6. **Oto A., Ernst R and GhulmiyyahL. (2009):** The role of MR cholan giopancreatography in the evaluation of pregnant patients with acute pancreaticobiliary disease. *Br J Radiol*., 82:279−285.
7. **Park DH., Kim MH and Lee SK. (2005):** Can MRCP replace the diagnostic role of ERCP for patients withch oledochalcysts? *Gastrointest Endosc*., 62:360−366.

8. **Shimotakahara A., Yamataka A and Yanai T. (2005):** Roux-en- Y hepaticojejunostomy or hepaticoduodenostomyfor biliary reconstruction during the surgical treatment of choledochal cyst: which is better? *Pediatr Surg Int*., 21:5−7.

9. **Singhavejsakul J and Ukarapol N. (2008):** Choledochal cysts: epidemiology and outcomes. *World Journal of Surgery*, 32:1385−1388.

10. **S?reide K., K?rner H and Havnen J. (2004):** Bileduct cysts in adults. *Br J Surg*., 91:1538−1548.

11. **Srimurthy KR and Ramesh S. (2006):** Laparoscopic management of pediatric choledochal cysts in developing countries: review of tencases. *Pediatr Surg Int*., 22:144−149.

12. **Todani T., Watanabe Y and Narusue M. (1977):** Congenital bileduct cysts: Classification, operative procedures, and review of thirtysevencases including cancer arising from choledochal cyst. *Am JSurg* ., 134:263−269.

13. **Todani T., Watanabe Y and Toki A. (2003):** Classification of congenitalbiliary cysticdisease: special reference to type Ic and IVa cysts with primary ductal stricture. *J Hepatobiliary Pancreat Surg*.,10:340-344.

14. **Woo R., LeD and AlbaneseCT. (2006):** Robot assisted laparoscopic resection of a type I choledochal cyst in a child. *J Laparoendosc Adv Surg Tech*., 16:179−183.

15. **Woon C., TanY and Oei CL. (2006):** Adult choledochal cysts: An audit of surgical treatment. *ANZ J Surg*., 76:981−986.

الجديد في مناجزة اكياس القنوات المرارية في البالغين

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تعد أكياس القنوات المرارية من الامراض الغير شائعة ، فمعدلات حدوثها تختلف إختلافا جوهريا في مناطق العالم ، وتعد قارة آسيا - وخاصة اليابان- من أكثر الماكن إنتشارا للمرض حيث يبلغ معدل الاصابة حالة لكل ألف حالة ، بينما يصل معدل الإصابة في الدول الغربية حالة لكل مئة وخمسين ألف حالة.

وأكياس القنوات المرارية عبارة عن تمددات في القنوات المرارية سواء كانت محدودة أو منتشرة إما داخل أو خارج الكبد ، وهي ناتجة غالبا عن عيوب خلقية في القنوات المرارية.

ويمثل التشخيص المبكر لأكياس القنوات المرارية تحديا للأطباء حيث يحتاج نسبة عالية من الشك عن طريق الثالوث الكلاسيكي : ألم متكرر بالجانب الايمن من البطن ، واليرقان ، وورم بالجانب الأيمن العلوي من البطن، ولكن مع ظهور الطرق الحديثة للتشخيص الإشعاعي بإستخدام الموجات فوق الصوتية والأشعة المقطعية والرنين المغناطيسي، ومنظار القنوات المرارية وأصبح التشخيص المبكر لأكياس القنوات المرارية أكثر سهولة.

ويعتبر التشخيص المبكر لتواجد أكياس بالقنوات المرارية من الضروريات لتلافي مضاعفاتها الخطيرة والتي تشمل الالتهابات المتكررة بالقنوات المرارية والبنكرياس ، وتكوين حصوات القنوات المرارية ، وتليف الكبد وإرتفاع ضغط الوريد البابي ، وتكوين أورام القنوات المرارية التي تتراوح معدلات حدوثها من 2,5 الي 26 %.

ويعتبر التدخل الجراحي هو السبيل الأمثل لعلاج القنوات المرارية والذي يشمل إستئصال الكيس المراري وتوصيل القناة الكبدية بالأمعاء والذي قد يصل في بعض الحالات الي إستئصال جزء من الكبد ، أو زراعة الكبد في الحالات المتأخرة.

ومن الجدير بالذكر أن متابعة الحالات بعد إجراء العملية أمراً مهماً للكشف المبكر عن تواجد أورام بالقنوات المرارية.