Choledochal cysts

Part 3 of 3: Management

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Much about the etiology, pathophysiology, natural course and optimal treatment of cystic disease of the biliary tree remains under debate. Gastroenterologists, surgeons and radiologists alike still strive to optimize their roles in the management of choledochal cysts. To that end, much has been written about this disease entity, and the purpose of this 3-part review is to organize the available literature and present the various theories currently argued by the experts. In part 3, we discuss the management of choledochal cysts, thus completing our comprehensive review.

Dans une large mesure, l'étiologie, la pathophysiologie, le cours naturel et le traitement optimal de la maladie kystique de l'arbre biliaire continuent de faire l'objet de débats. Les gastroentérologues, les chirurgiens et les radiologues cherchent toujours à optimiser leur rôle respectif dans la prise en charge des kystes du cholédoque. C'est pourquoi les chercheurs ont beaucoup écrit à propos de cette entité morbide, et le présent examen en trois parties a pour objet d'organiser les études publiées et de présenter les diverses théories que font actuellement valoir les experts. Dans la troisième partie, nous discutons de la prise en charge des kystes du cholédoque, ce qui complète notre examen détaillé.

Choledochal cysts (CCs) are single or multiple dilatations of the intrahepatic or extrahepatic biliary tree. If left untreated, they can cause morbidity and mortality from recurrent cholangitis, pancreatitis, sepsis, liver abscesses and cholangiocarcinoma. Comprehensive treatment involves medical management of complications, surgery and long-term follow-up. The first 2 parts of this 3-part series reviewed the background of CCs and explained how to accurately diagnose this disease. The final article in this series discusses the management of CC, from historical treatment to current standards of practice, and includes special considerations such as pregnancy.

Management

Management of CC depends on the type of cyst. Treatment of types I and IV A cysts has undergone much change in the past years. Although Mcwhorter first described cyst excision and hepaticojejunostomy in 1924, this surgery was initially abandoned because of multiple complications.[1] Surgical strategies of cyst marsupialization and choledochoraphy failed because of significant mortality and morbidity.[1,2]

Subsequently, internal drainage of cysts via cystenterostomy became popular. Depending on anatomic proximity, cysts were incised and anastomosed to the duodenum or jejunum.[2] Although this operation resulted in periprocedural relief of symptoms, multiple complications resulted. Reflux of the enteric contents into the cyst and biliary tree resulted in recurrent ascending cholangitis. The site of anastomosis was also prone to stricture formation, resulting in obstruction, bile stasis, stone formation and recurrent cholangitis. Most importantly, surgeons found that leaving the cyst intact carried a significant risk of malignant transformation.[3,4] The overall success rate of internal drainage procedures is 30%, the risk of postoperative malignancy is 30%, the

mortality rate is 11%, and more than half who undergo this procedure require re-operation.[5,6] Therefore, internal drainage is currently thought to be a dangerous and incomplete treatment of CC.[5]

Instead, surgeons favour complete cyst excision and hepaticoenterostomy.[7,8] This separates the biliary tree from the pancreatic duct, thus ending the mixing of pancreatic and biliary secretions thought to be responsible for the pathogenesis of the disease; it also excises the damaged and presumably premalignant cyst tissue.[9] If left in situ, the risk of cancer in the retained cyst is as high as 50% and occurs 15 years earlier than primary cancer.[9,10] Therefore, the cyst should be excised completely from the hepatic hilum to the pancreatic duct.[8,10,11] Recurrent inflammation, cholangitis and pancreatitis result in fibrosis of the ducts and adhesion to surrounding structures, making excision difficult. Intramural saline injection may separate the dissection planes and facilitate excision.[12] If the cyst cannot be completely excised, the mucosa should either be stripped or destroyed by abrasion and iodine or alcohol application.[5,12] Any patient with a remnant cyst should receive regular surveillance via ultrasound.[5] The hepaticoenterostomy can either be a hepaticoduodenostomy or a Roux-en-Y hepaticojejunostomy (RYHJ). The success rate of RYHJ has been shown to be as high as 92%.[5] This procedure has a reported complication rate of 7%, compared with a complication rate of 42% with hepaticoduodenostomy.[13] Hepaticoduodenostomy carries with it the risk of bilious gastric reflux, gastritis and esophagitis, ulceration and malignant disease.[4,13,14] Furthermore, Todani and colleagues[15] reverted from advocating hepaticoduodenostomy as the procedure of choice when they discovered a patient with hilar adenocarcinoma after excision. They hypothesize that that the reflux of bile and active pancreatic enzymes from the duodenum can irritate the hilar epithelium and lead to malignant transformation.[13,15] Many surgeons recommend end-to-end RYHJ to avoid the formation of a long blind pouch, which can result in bile stasis, reflux, cholangitis and stone formation.[16–19] Authors also recommend creating a wide stoma at the hepatic hilum by extending the incisions up the lateral walls of the hepatic ducts to allow free drainage and avoid anastomotic stricture.[4,9,17] The minimum diameter of the stoma has been suggested to be 3 cm.[5] After cyst excision and hepaticoenterostomy, patients symptoms improve, intrahepatic duct dilations decompress and hepatic fibrosis and varices regress.[20,21] The complications of cystenterostomy and benefits of cyst excision and hepaticoenterostomy are both so substantial that surgeons now recommend revision of previous internal drainage procedures even for patients with no symptoms or complications.[9,22]

Early complications of cyst excision and hepaticoenterostomy include anastomotic leak, pancreatic leak with injury to the pancreatic duct, bowel obstruction due to intussusception, and bowel kinking due to manipulation or adhesions. Late complications include peptic ulcer disease, cholangitis, biliary calculi, pancreatitis, liver failure and cancer.[9,23,24] Fibrosis and inflammation of cyst tissue at the time of surgery, such that the anastomosed margins are friable, result in poor healing, leakage and anastomotic stricture. Because fibrosis and inflammation increase with age, surgical complications become more common with older age at surgery, and surgery should be done as early as possible.[16,17] Cholangitis and calculi usually occur as a result of anastomotic stricture leading to bile stasis.[16]

Both cholangitis and pancreatitis can also result from duct stenosis or obstruction from debris, lithiasis and protein plugs. Pancreatic remnant cysts often cause these obstructive factors.[25] Many surgeons have had success and report a dramatic decrease in the complication rate with intraoperative cystendoscopy with identification and correction of stenosis and wash out of stones, debris and plugs.[6,16,26,27] Cystendoscopy is also useful to identify the location of the pancreatic duct so that as much of the distal common bile duct can be excised without pancreatic injury.[28] If intrahepatic duct stenoses are so high that they cannot be reached, a hepaticocutaneous jejunostomy for continued balloon dilation and stone extraction may be warranted.[15] Many authors also suggest the use of perioperative and long-term antibiotics to minimize the incidence of cholangitis.[29]

Postexcisional malignant disease, which has an incidence of 0.7%–6%, is thought to be due to remnant cyst tissue or subclinical malignant disease not detected before surgery.[4,30] Therefore, some authors recommend intraoperative endoscopic ultrasonography and pathology of frozen sections to rule out dysplasia, hyperplasia and malignant disease.[12,30] All patients with CC also require life-long follow-up for cancer, usually via serial ultrasonography and monitoring of liver enzymes.[11,31,32]

Surgery may be hindered by cirrhosis, portal hypertension and varices. Large pericystic varices, especially in the hepaticoduodenal ligament, increase the risk of postoperative bleeding. Therefore, if there is clinical or radiological evidence of cirrhosis or portal hypertension, esophagogastroscopy should be performed to identify and assess the extent of the varices. If large pericystic varices are a concern, portosystemic shunting can be performed for decompression before surgery.[13,21] Gallstone ileus after hepaticoenterostomy has also been reported; this operation facilitates the passage of stones into the enteric tract.[33]

Recently, many authors have reported success performing cyst excision and RYHJ via laparoscopy, with quicker recovery (mean hospital stay 5.5 days), less adhesions and improved cosmesis and ease of surgery because of magnification of the operative field.[29,34–42] Although initial laparoscopic surgeries took 9–10 hours, technological advances and operator experience have shortened this to a reasonable 4.5–5.5 hours.[38–40] With the advent of robot-assisted surgery, which has improved manual dexterity, this operative time

continues to become shorter.[42] Laparoscopic treatment of CC is still evolving and promises many future benefits.

Various modifications to the surgical treatment described have been proposed. Shah and Shah[43] proposed an appendiceal conduit. In this surgery, the cecum is mobilized to the splenic flexure, the appendix and its vascular pedicle are dissected, the cecal end is anastomosed to the hepatic duct, and the distal end is incised and then anastomosed to the jejunum as a tubular structure. Although the authors argue that this procedure is superior because of reduced risk of cholangitis from a high appendiceal lymphoid follicle content and physiologic duodenal bile drainage, this procedure has not gained popularity.[43,44] Chang described a procedure in which a spur valve was placed in the ascending limb of the RYHJ in an attempt to prevent biliary reflux and cholangitis, but this resulted in a reoperation rate as high as 15% and complications of recurrent cholangitis and obstructive jaundice.[45,46] Raffensperger[47] and Zhang[48] proposed the Chicago–Beijing procedure, which is still commonly used in China. This technique comprises cyst excision with a jejunal conduit between the hepatic stump and the duodenum, with an antireflux spur valve at the jejunoduodenal anastomosis.[45,47–49] The benefits of this type of surgery are physiologic bile drainage and a valve that prevents reflux, whereas the disadvantages are a long, complicated procedure and anastomotic stricture formation. Surgeons performing this procedure report a low reoperation rate of 0.8% in a large number of patients (n = 481).[45] Other surgeons have abandoned this procedure, however, because of a high incidence of postoperative pain, which has been attributed to reflux biliary gastritis.[12] Although all of these innovations appear great in theory, actual benefit has not yet been demonstrated, and cyst excision and RYHJ remain the procedures of choice.

Many patients are diagnosed with CC while acutely ill with active cholangitis, pancreatitis or rupture and bile peritonitis. In these conditions, the patient's physical state and intra-abdominal inflammation make the risks of surgery substantial. Furthermore, operating on acutely inflamed tissue results in poor healing, scarring and anastomotic stricture. Therefore, temporary measures should be performed, with definitive surgery performed when the patient's clinical condition allows.[50] For active cholangitis and pancreatitis, the procedure of choice is external drainage via T-tube or percutaneous hepaticostomy.[5,51,52] This is preferred over internal drainage because the only scarring is around the percutaneous tract, whereas the latter results in fibrosis of the duodenum, pericystic vascular structures and the hepaticoduodenal ligament, all of which make subsequent surgery very difficult.[20] Cyst rupture should include laparotomy and washout of the bile, external drainage and antibiotics for stabilization before definitive surgery.[2,9,51,53,54]

The risk of malignant disease with type II and III cysts is exceedingly low, and, thus, complete excision is not necessary. Simple excision of type II cysts is sufficient.[12,41] Choledochoceles often just require endoscopic sphincterotomy to allow free duodenal drainage of bile and stones.[55–57] Given the possibility that pancreatic and biliary secretions can mix within the choledochocele and create a precancerous state, some authors recommend sphincterotomy even in asymptomatic patients.[56,58] Endoscopic excision via snare cautery is also possible for small cysts.[55] Some surgeons believe that the common bile duct and pancreatic duct should be separated and reanastomosed to the duodenum to prevent pancreaticobiliary mixing.[12] Large choledochoceles may cause biliary, duodenal or gastric outlet obstruction, in which case duodenotomy and cyst excision is warranted.[57,59]

Forme fruste CC are associated with significant risk of bile duct and gall bladder cancer, and treatment of this condition requires at least cholecystectomy.[12] However, many authors believe that this is not enough to prevent malignant disease and advocate excision of the choledochus and hepaticoenterostomy.[12,60]

Treatment of type IV A and V disease remains difficult. Type IV A is treated by cyst excision and a wide hilar hepaticoenterostomy, but patients often continue to have symptoms because of intrahepatic disease. If the intrahepatic involvement is localized, a segmental hepatectomy may be performed.[5,61,62] For diffuse disease, a percutaneous hepaticojejunostomy may allow for continuous stone extraction and dilation.[6,12,62–64] Surgical or endoscopic unroofing of some intrahepatic cysts can also be performed for bile drainage.[65] Similarly, localized Caroli disease may be treated by hepatic lobectomy.[13,65–67] Diffuse disease with recurrent or life-threatening cholangitis, liver failure, cirrhosis and portal hypertension or malignant disease requires orthotopic liver transplantation.[11,12,65,67–72] Some authors recommend early liver transplant if possible, because prognosis is very poor once malignant disease develops.[70] Although prophylactic transplant is not warranted, aggressive surveillance for malignant disease in asymptomatic or minimally symptomatic patients is required.[11,69] Complications of transplant include bleeding, sepsis, hepatic artery thrombosis and rejection.[65,73]

Recurrent lithiasis and cholangitis in both type IV A and V cysts can also be conservatively treated with prophylactic antibiotics for patients who are well, and intravenous and intraductal antibiotics can be used for ill patients; endoscopic or percutaneous lithotripsy and ursodeoxycholic acid can also be used.[16,65,66,74–77] Ursodeoxycholic acid has proven effective in dissolving pre-existing stones and preventing the formation of new stones.[78,79]

Malignant disease within the biliary tree mandates excision of the extrahepatic bile duct and adjacent liver, with regional lymph node excision.[3,10,80] Unfortunately, less than 10% of cancers are resectable at diagnosis.[81] Metastatic disease that affects the surrounding vasculature, organs or peritoneum may need percutaneous, endoscopic or surgical

bile duct stent placement.[3] A prophylactic or therapeutic gastroenterostomy to bypass the affected enteric tract and relieve obstruction may also be necessary.[12] Distal malignant disease within the pancreatic head requires a Whipple procedure.[82] Adjuvant chemotherapy or radiotherapy or both may increase survival, although prognosis after diagnosis of cancer is very poor.[80,81,83]

Many previously asymptomatic women present during pregnancy for a number of reasons, including obstruction of the cyst by the gravid uterus, further stasis of pancreaticobiliary secretions because of biliary hypomotility, and cyst rupture because of increased intra-abdominal pressure during pregnancy and labour.[84,85] Presenting symptoms are usually abdominal pain, fever and vomiting, usually due to cholangitis or pancreatitis.[84–88] Diagnosis by ultrasonography may be difficult because of obscuration and alteration of normal anatomy by the gravid uterus.[84] Given that computed tomography scans expose the fetus to ionizing radiation, magnetic resonance imaging (MRI) has been recommended as the imaging modality of choice.[84,88] Management of CC during pregnancy is difficult because of the surgical risk to both mother and fetus. Incidentally found CC should be followed with serial ultrasonography, and symptoms or rapid cyst enlargement should be treated conservatively.[85] Patients with active cholangitis or pancreatitis should also receive conservative treatment of hospital admission and close observation, external drainage and antibiotics. Despite the label "conservative treatment," nonsurgical management should be aggressive because pancreatitis carries a maternal mortality rate of 20% and a fetal mortality rate of 38%.[87] High intra-abdominal pressure during labour may cause cyst rupture, and many surgeons recommend elective caesarian section in the third trimester.[84,85,87,88] Subsequently, definitive cyst excision and hepaticoenterostomy should be performed after delivery.[84,85,87,88] Cyst rupture may mandate emergent surgery for bile evacuation and washout, but this should be followed with external drainage, and definitive surgery should be performed during the postpartum period.[85]

Ultrasound and MRI have been used to antenatally diagnose CC, even before the onset of signs or symptoms.[89–92] Histopathology shows increased incidence and grade of liver fibrosis in pediatric patients with increasing age at surgery.[93–95] Such fibrosis has been shown to regress after surgery.[94] Furthermore, the longer surgery is delayed, the greater the potential for complications such as cyst rupture.[63] Serial ultrasonography shows rapid cyst enlargement after birth at a rate of 2 mm per week, perhaps because of increased pancreatic and biliary secretion after the initiation of feedings.[95–97] Additionally, the longer the biliary tree is exposed to the chronic inflammation associated with CC, the greater the risk of malignant transformation.[93] Finally, the surgical complication rate is almost negligible in the neonatal period but increases with age at surgery.[23] For all of these reasons, most pediatric surgeons advocate neonatal cyst excision for prenatally diagnosed CC, even before the onset of symptoms.[93,95–97] While waiting for surgery, neonatal patients should receive serial ultrasonography and liver enzyme measurements; a rapidly enlarging cyst, cholangitis or worsening liver function should prompt expedient surgery.[57,94,97,98]

CONCLUSION

Although rare, biliary cystic disease remains an interesting clinical problem. The current well-established classification system is being challenged by the notion that these subtypes may be many different disease entities within the same anatomic location. The recurrent cholangitis, pancreatitis and malignant diseases associated with choledochal cysts mandates early diagnosis via ultrasonography and cholangiography and definitive surgical management. Surgeons currently agree that all patients require complete cyst excision and hepaticoenterostomy. In this 3 part review series, we attempted to comprehensively describe all aspects of this disease to enhance clinical understanding, explain current debates and spark further research.

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References

1. Kasai M, Asakura Y, Tamia Y. Surgical treatment of choledochal cyst. *Ann Surg* 1970;172:844-51.
2. Daniel DS. Choledochal cyst: report of a case. *Ann Surg* 1962;155: 902-5.
3. Tsuchiya R, Harada N, Ito T, et al. Malignant tumors in choledochal cysts. *Ann Surg* 1977;186:22-8.
4. Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. *J Hepatobiliary Pancreat Surg* 1999;6: 207-12.
5. Tao KS, Lu YG, Wang T, et al. Procedure for congenital choledochal cysts and curative effect analysis in adults. *Hepatobiliary Pancreat Dis Int* 2002;1:442-5.
6. Saing H, Han H, Chen KL. Early and late results of excision of choledochal cyst. *J Pediatr Surg* 1997;32:1563-6.
7. Gardikis S, Antypas S, Kambouri K, et al. The Roux-en-Y procedure in congenital hepato-biliary disorders. *Rom J Gastroenterol* 2005;14: 135-40.
8. Yoshikane H, Hashimoto S, Hidano H. Multiple early bile duct carcinoma associated with congenital choledochal cyst. *J Gastroenterol* 1998;33:454-7.
9. Todani T, Watanabe Y, Toki A, et al. Reoperation for congenital choledochal cyst. *Ann Surg* 1988;207:142-7.
10. Fieber SS, Nance FC. Choledochal cyst and neoplasm: a comprehensive review of 106 cases and presentation of two original cases. *Am Surg* 1997;63:982-7.
11. Bismuth H, Krissat J. Choledochal cystic malignancies. *Ann Oncol* 1999;10(Suppl 4):94-8.

12. Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. *J Hepatobiliary Pancreat Surg* 2003;10:352-9.
13. Shimotakahara A, Yamataka A, Yanai T. Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy for biliary reconstruction during the surgical treatment of choledochal cyst: Which is better? *Pediatr Surg Int* 2005;21:5-7.
14. Takada H, Hamada Y, Watanabe K. Duodenogastric reflux following biliary reconstruction after excision of choledochal cyst. *Pediatr Surg Int* 2005;21:1-4.
15. Todani T, Watanabe Y, Urushihara N, et al. Biliary complications after excisional procedure for choledochal cyst. *J Pediatr Surg* 1995; 30:478-81.
16. Yamataka A, Ohshiro K, Okada Y, et al. Complications after cyst excision with hepaticoenterostomy for choledochal cysts and their surgical management in children versus adults. *J Pediatr Surg* 1997; 32:1097-102.
17. Miyano T, Yamataka A, Kato Y, et al. Hepaticoenterostomy after excision of choledochal cyst in children: a 30-year experience with 180 cases. *J Pediatr Surg* 1996;31:1417-21.
18. Yamataka A, Kobayashi H, Shimotakahara A, et al. Recommendations for preventing complications related to Roux-en-Y hepaticojejunostomy performed during excision of choledochal cyst in children. *J Pediatr Surg* 2003;38:1830-2.
19. Ajiki T, Suzuki Y, Okazaki T, et al. A large stone detected in Roux-en-Y jejunal limb 20 years after excision of congenital choledochal cyst. *Surgery* 2006;139:129-30.
20. Chaudhary A, Dhar P, Sachdev A. Reoperative surgery for choledochal cysts. *Br J Surg* 1997;84:781-4.
21. Gananadha S, Smith RC. Hepatobiliary and pancreatic: anomalous pancreatobiliary junction with choledochal cyst. *J Gastroenterol Hepatol* 2006;21:776.
22. de Vries JS, de Vries S, Aronson DC, et al. Choledochal cysts: age of presentation, symptoms, and late complications related to Todani's classification. *J Pediatr Surg* 2002;37:1568-73.
23. Lee SC, Kim HY, Jung SE, et al. Is excision of a choledochal cyst in the neonatal period necessary? *J Pediatr Surg* 2006;41:1984-6.
24. Li MJ, Feng JX, Jin FQ. Early complications after excision with hepaticoenterostomy for infants and children with choledochal cyst. *Hepatobiliary Pancreat Dis Int* 2002;1:281-4.
25. Koshinaga T, Hoshino M, Inoue M, et al. Pancreatitis complicated with dilated choledochal remnant after congenital choledochal cyst excision. *Pediatr Surg Int* 2005;21:936-8.
26. Yamataka A, Segawa O, Kobayashi H, et al. Intraoperative pancreatoscopy for pancreatic duct stone debris distal to the common channel in choledochal cyst. *J Pediatr Surg* 2000;35:1-4.
27. Shimotakahara A, Yamataka A, Kobayashi H, et al. Massive debris in the intrahepatic bile ducts in choledochal cyst: possible cause of postoperative stone formation. *Pediatr Surg Int* 2004;20:67-9.
28. Miyano T, Yamataka A, Kato Y, et al. Choledochal cysts: special emphasis on the usefulness of intraoperative endoscopy. *J Pediatr Surg* 1995;30:482-4.
29. Ure BM, Schier F, Schmidt AI. Laparoscopic resection of congenital choledochal cyst, choledochojejunostomy, and extraabdominal Roux-en-Y anastomosis. *Surg Endosc* 2005;19:1055-7.
30. Kobayashi S, Asano T, Yamasaki M. Risk of bile duct carcinogenesis after excision of extrahepatic bile ducts inpancreaticobiliary maljunction. *Surgery* 1999;126:939-44.
31. Franko J, Nussbaum ML, Morris JB. Choledochal cyst cholangiocarcinoma arising from adenoma: case report and a review of the literature. *Curr Surg* 2006;63:281-4.
32. Di Sena V, de Paulo GA, Macedo EP, et al. Choledochal cyst mimicking a pancreatic pseudocyst: case report and review. *Gastrointest Endosc* 2003;58:620-4.
33. Gaiani S, Serra C, Cervellera M, et al. Gallstone ileus in Caroli's disease. *Am J Gastroenterol* 2000;95:3642-3.
34. Shimura H, Tanaka M, Shimizu S, et al. Laparoscopic treatment of congenital choledochal cyst. *Surg Endosc* 1998;12:1268-71.
35. Srimurthy KR, Ramesh S. Laparoscopic management of pediatric choledochal cysts in developing countries: review of ten cases. *Pediatr Surg Int* 2006;22:144-9.
36. Lee H, Hirose S, Bratton B, et al. Initial experience with complex laparoscopic biliary surgery in shildren: biliary atresia and choledochal cyst. *J Pediatr Surg* 2004;39:804-7.
37. Tan HL, Shankar KR, Ford WDA. Laparoscopic resection of type I choledochal cyst: report of two cases in children. *Surg Endosc* 2003; 17:1495.
38. Chowbey PK, Katrak MP, Sharma A, et al. Complete laparoscopic management of choledochal cyst: report of two cases. *J Laparoendosc Adv Surg Tech A* 2002;12:217-21.
39. Le DM, Woo RK, Sylvester K, et al. Laparoscopic resection of type 1 choledochal cysts in pediatric patients. *Surg Endosc* 2006;20:249-51.
40. Watanabe Y, Sato M, Takui K, et al. Laparoscopic-assisted minimally invasive treatment for choledochal cyst. *J Laparoendosc Adv Surg Tech A* 1999;9:415-8.
41. Liu DC, Rodriguez JA, Meric F, et al. Laparoscopic excision of a rare type II choledochal cyst: case report and review of the literature. *J Pediatr Surg* 2000;35:1117-9.
42. Tanaka M, Shimizu S, Yokohata K, et al. Laparoscopically assisted resection of choledochal cyst and Roux-en-Y reconstruction. *Surg Endosc* 2001;15:545-51.
43. Shah AA, Shah AV. Appendix as a biliary conduit for choledochal cyst in children. *Eur J Pediatr Surg* 2005;15:128-31.
44. Wei MF, Qi BQ, Xia GL, et al. Use of appendix to replace the choledochus. *Pediatr Surg Int* 1998;13:494-6.
45. Fu M, Wang YX, Zhang JZ. Evolution in the treatment of choledochus cyst. *J Pediatr Surg* 2000;35:1344-7.
46. Chang CC. Antireflux spur valve in cholenterostomy. *Jpn Soc J Pediatr Surg* 1982;18:73-5.
47. Raffensperger JG. *Swenson's pediatric surgery*. 5th ed. Norwalk (CT): Appleton & Lange; 1995.
48. Zhang JZ, Wang YX, Wei LQ, et al. The spur valve jejunal interposition in choledochus cystectomy. *Chin Med J (Engl)* 1987;100:535-40.
49. Narasimha Rao KL, Mitra SK, Kochher R, et al. Jejunal interposition hepaticoduodenostomy for choledochal cyst. *Am J Gastroenterol* 1987; 82:1042-5.
50. Woon CYL, Tan YM, Oei CL. Adults choledochal cysts: an audit of surgical management. *ANZ J Surg* 2006;76:981-6.
51. Lee CC, Levine DA, Tunik MG, et al. A case report: type I choledochal cyst induced pancreatitis in a 15-month-old child. *Pediatr Emerg Care* 2000;16:265-7.
52. Gulati MS, Srivastava DN, Paul SB, et al. Pre-operative management of congenital choledochal cyst with ultrasound-guided percutaneous choledochalcystostomy. *Australas Radiol* 1999;43:514-6.
53. Kiresi DA, Karabacakoglu A, Dilsiz A, et al. Spontaneous rupture of choledochal cyst presenting in childhood. *Turk J Pediatr* 2005;47:283-6.
54. Maheshwari M, Parekh BR, Lahoti BK. Biliary peritonitis: a rare presentation of perforated choledochal cyst. *Indian Pediatr* 2002;39:588-92.
55. Chatila R, Andersen DK, Topazian M. Endoscopic resection of a choledochocele. *Gastrointest Endosc* 1999;50:578-81.

56. Horaguchi J, Fujita N, Kobayashi G. Clinical study of choledochocele: Is it a risk factor for biliary malignancies? *J Gastroenterol* 2005;40:396-401.
57. Masetti R, Antinori A, Coppola R. Choledochocele: changing trends in diagnosis and management. *Surg Today* 1996;26:281-5.
58. Ohtsuka T, Inoue K, Ohuchida J. Carcinoma arising in choledochocele. *Endoscopy* 2001;33:614-9.
59. Ramos A, Castell J, Pinto I. Intestinal intussusception as a presenting feature of choledochocele. *Gastrointest Radiol* 1990;15:211-4.
60. Miyano G, Yamataka A, Shimotakahara A. Cholecystectomy alone is inadequate for treatment of form fruste choledochal cyst: evidence from a rare but important case. *Pediatr Surg Int* 2005;21:61-3.
61. Hussain ZH, Bloom DA, Tolia V. Caroli's disease diagnosed in a child by MRCP. *Clin Imaging* 2000;24:289-91.
62. Lal R, Agarwal S, Shivhare R, et al. Type IV-A choledochal cysts: a challenge. *J Hepatobiliary Pancreat Surg* 2005;12:129-34.
63. Kirimlioglu V, Yilmaz S, Katz DA, et al. Choledochal cyst spontaneously rupturing the hepatic artery. *Dig Dis Sci* 2000;45:544-8.
64. Hewitt PM, Krige JEJ, Bornman PC, et al. Choledochal cysts in adults. *Br J Surg* 1995;82:382-5.
65. Habib S, Shakil O, Couto OF, et al. Caroli's disease and orthotopic liver transplantation. *Liver Transpl* 2006;12:416-21.
66. Izawa K, Tanaka K, Furui J, et al. Extended right lobectomy for Caroli's disease: report of a case and review of hepatectomized cases in Japan. *Surg Today* 1993;23:649-55.
67. Vlachogiannakos J, Potamianos S, Triantos C. Monolobar Caroli's disease complicated by cholangiocarcinoma in a 70-year-old man, previously asymptomatic. *Gastrointest Endosc* 2004;60:297-300.
68. Takatsuki M, Uemoto S, Inomata Y, et al. Living-donor liver transplantation for Caroli's disease with intrahepatic adenocarcinoma. *J Hepatobiliary Pancreat Surg* 2001;8:284-6.
69. Bockhorn M, Malagó M, Lang H, et al. The role of surgery in Caroli's disease. *J Am Coll Surg* 2006;202:928-32.
70. Schiano TD, Fiel MI, Miller CM, et al. Adult presentation of Caroli's syndrome treated with orthotopic liver transplantation. *Am J Gastroenterol* 1997;92:1938-40.
71. Levy AD, Rohrman CA, Murukata LA, et al. Caroli's disease: radiologic spectrum with pathologic correlation. *AJR Am J Roentgenol* 2002;179:1053-7.
72. Sans M, Rimola A, Navasa M. Liver transplantation in patients with Caroli's disease and recurrent cholangitis. *Transpl Int* 1997;10:241-4.
73. De Kerckhove L, De Meyer M, Verbaandert C, et al. The place of liver transplantation in Caroli's disease and syndrome. *Transpl Int* 2006;19:381-8.
74. Karim AS. Caroli's disease. *Indian Pediatr* 2004;41:848-50.
75. Pinto RB, Lima JP, da Silveira TR, et al. Caroli's disease: report of 10 cases in children and adolescents in southern Brazil. *J Pediatr Surg* 1998;33:1531-5.
76. Harjai MM, Lal BK. Caroli disease. *Pediatr Surg Int* 2000;16:431-2.
77. Wu KL, Changchien CS, Kuo CM, et al. Caroli's disease — a report of two siblings. *Eur J Gastroenterol Hepatol* 2002;14:1397-9.
78. Ros E, Navarro S. Ursodeoxycholic acid treatment of primary hepatolithiasis in Caroli's syndrome. *Lancet* 1993;342:404-6.
79. Naga MI, Suleiman DN. Endoscopic management of choledochal cyst. *Gastrointest Endosc* 2004;59:427-32.
80. Harris M, Angus P, Davis ID. Choledochal cyst and squamous-cell carcinoma of the biliary tract. *Intern Med J* 2002;32:491.
81. Benjamin IS. Biliary cystic disease: the risk of cancer. *J Hepatobiliary Pancreat Surg* 2003;10:335-9.
82. Jordan PH, Goss JA, Rosenberg WR, et al. Some considerations for management of choledochal cysts. *Am J Surg* 2004;187:790-5.
83. Patel YD, Sarwar U, Morehouse HT, et al. Radiological management of an inoperable cholangiocarcinoma arising from a choledochal cyst. *Clin Radiol* 2001;56:503-19.
84. Palled SR, Doddihal HM, Mahantshetty UM. Choledochal cyst carcinoma treated with stereotactic radiotherapy. *Clin Oncol (R Coll Radiol)* 2006;18:153-4.
85. Wu DQ, Zheng LX, Wang QS, et al. Choledochal cysts in pregnancy: case management and literature review. *World J Gastroenterol* 2004;10:3065-9.
86. Nasu K, Matsuki S, Kawano Y. Choledochal cyst diagnosed and conservatively treated during pregnancy. *Am J Perinatol* 2004;21:463-8.
87. Adair CD, Castillo R. Caroli's disease complicating pregnancy. *South Med J* 1995;88:763-4.
88. Beattie GJ, Keay S, Muir BB, et al. Acute pancreatitis with pseudocyst formation complicating pregnancy in a patient with a co-existent choledochal cyst. *Br J Obstet Gynaecol* 1993;100:957-9.
89. Wig JD, Goenka MK, Chawla YK, et al. Cholangitis secondary to choledochal cyst in pregnancy and puerperium. *J Clin Gastroenterol* 1997;25:489-91.
90. Wong AMC, Cheung YC, Liu YH, et al. Prenatal diagnosis of choledochal cyst using magnetic resonance imaging: a case report. *World J Gastroenterol* 2005;11:5082-3.
91. Chen CP, Cheng SJ, Sheu JC, et al. Third-trimester evaluation of choledochal cyst using magnetic resonance imaging. *Prenat Diagn* 2004;24:838-9.
92. Chen CP, Cheng SJ, Chang TY, et al. Prenatal diagnosis of choledochal cyst using ultrasound and magnetic resonance imaging. *Ultrasound Obstet Gynecol* 2004;23:93-5.
93. Gallivan EK, Crombleholme TM, D'Alton ME. Early prenatal diagnosis of choledochal cyst. *Prenat Diagn* 1996;16:934-7.
94. Lugo-Vicente HL. Prenatally diagnosed choledochal cysts: Observation or early surgery? *J Pediatr Surg* 1999;30:1288-290.
95. Suita S, Shone K, Kinugasa Y, et al. Influence of age on the presentation and outcome of choledochal cyst. *J Pediatr Surg* 1999;34:1765-8.
96. Siddiqui MMF, Grier D, Cusick E. Postnatal rupture of an antenatally diagnosed choledochal cyst: first case report. *Acta Paediatr* 2006; 95:115-7.
97. Hamada Y, Tanano A, Sato M, et al. Rapid enlargement of a choledochal cyst: antenatal diagnosis and delayed primary excision. *Pediatr Surg Int* 1998;1998:419-21.
98. Clifton MS, Goldstein RB, Slavotinek A, et al. Prenatal diagnosis of familial type I choledochal cyst. *Pediatrics* 2006;117:e596-600.