

Partial Hepatectomy and Total Cyst Excision is Curative for Localized Type IV-A Biliary Duct Cysts – Report of Four Cases and Review of Management

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Key words

- type IV-A choledochal cyst
- uniductal/localized variety
- hepatectomy
- curative resection
- children

Abstract



Purpose: With the increasing use of newer modalities such as CT cholangiography and MRCP offering an accurate delineation of the ductal anatomy of choledochal cysts, the incidence of type IV-A biliary duct cysts has increased. Although the management of the more common type I cysts is well established, that of type IV-A cysts is still controversial. The localized/unilobar variety of type IV-A cysts is a unique entity amenable to curative surgical management.

Methods: Between Jan 2000 and Jan 2005, 10 of 25 cases with choledochal cysts were diagnosed as having type IV- cysts. Four of these were localized or unilobar variants affecting only one lobe of the liver. Three cases had a left ductal involvement and one had a right ductal involvement of their intrahepatic component. We describe the presentation and curative surgical management of these patients with a unilobar variety of type IV-A cysts and discuss the management options for type IV-A cysts in the literature.

Results: One child with previous surgical treatment for presumed type 1 cyst, diagnosed as having a type IV-A right ductal cyst complicated with liver abscess, underwent right hepatectomy, two children had a left hepatectomy and one child had a left lateral lobectomy. All four children had uncomplicated intraoperative and postoperative courses. After 4-8 years' follow-up, all are symptom-free. There were no biliary tract or hepatic abnormalities on radiological surveillance with MRCP.

Conclusions: In some series, the incidence of type IV-A choledochal cysts is equal to or higher than that of type 1 cysts. MRCP and intraoperative cholangiogram can help to confirm the anatomical varieties and associated ductal strictures. In the unilobar/uniductal varieties, complete cyst excision can be safely achieved in children through an additional hepatectomy/lobectomy. This can result in a curative status, leaving the child free from complications such as cholangitis, abscess, hepatolithiasis and the risk of biliary duct malignancy.

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Introduction



Congenitally dilated extrahepatic and intrahepatic bile ducts have been classified as type IV-A choledochal cysts by Todani et al. [13] The use of modalities such as cholangiogram, ERCP, CT scan and MRCP has led to accurate anatomical diagnoses of various types of choledochal cyst. In recent series the incidence of type IV-A cysts has been reported to be between 33–45% [3, 6, 9, 10, 18]. The management of such cysts has also undergone a sea change in the last 3 decades. Internal drainage procedures have been abandoned in favor of excision of extrahepatic cysts and wide biliary enteric anastomosis of the common hepatic duct. There is now widespread acceptance that the preferred treatment for type

I cysts is surgical excision with Roux-en-Y biliary reconstruction to the confluence of the hepatic ducts [3,9]. This eliminates biliary stasis and recurrent cholangitis related to the extrahepatic abnormality and removes the major site of susceptibility to malignancy. However, anatomical variations (localized vs. bilobar intrahepatic involvement), associated bile duct stricture, pathological complications of intrahepatic cysts (cholangitis, hepatic abscess, rupture and intrahepatic calculi), and issues of malignancy in the residual cysts mean that the optimum surgical cure for type IV cysts still controversially discussed.

In an attempt to achieve complete eradication of cysts while preventing complications, a few authors have performed partial hepatectomies

for a localized form of Caroli's disease [18] and type IV-A cysts [3,6,10] in adults. We present 4 cases of type IV-A biliary duct cysts in children with selective involvement of left intrahepatic duct system in 3 and the right intrahepatic duct system in one (unilobar form of type IV-A). We present our management of these cases with intent of complete surgical cure and discuss the management issues relating to multiple biliary duct cysts (IV-A) presented in the literature.

Material and Methods

Records of cases diagnosed and treated for choledochal cysts between Jan 2000 and Jan 2005 were reviewed retrospectively. Only patients diagnosed as having type IV-A disease, specifically with uniductal intrahepatic involvement, were included in the review. Records of clinical history, preoperative investigations including liver function test, ultrasound, CT/MRCP, preoperative classification of the cyst, intraoperative cholangiography, operative procedure and immediate postoperative recovery and any complications were analyzed. Follow-up records were reviewed for any symptoms suggestive of cholangitis, the status of liver function tests and the radiological assessment of the liver and biliary duct system by MRCP/CT to rule out any dilatation, calculus and neoplasia.

Results

Between Jan 2000 and Jan 2005, a total of 25 children with choledochal cysts were treated by the authors; out of these a

total of 10 children were diagnosed as having type IV-A cysts, 4 of whom presented with uniductal or localized intrahepatic involvement. The accounts of these 4 cases are given below.

Case 1

A 7-year-old boy was previously diagnosed as having type IV-A choledochal cyst and underwent excision of an extrahepatic cyst with wide hilar hepaticojejunostomy at 4 years of age. He presented with recurrent episodes of febrile illness, associated right hypochondrial pain, vomiting and jaundice requiring hospitalization. He had been treated for cholangitis by i.v. fluids and antibiotics at another center. Upon evaluation, he was noticed to be febrile (38.7°C) and mildly jaundiced (total bilirubin 2.5 mg/dl); USG revealed a right lobar cystic space-occupying lesion with echogenic content measuring 8×6 cm², and CT scan showed features of hepatic abscess with right ductal cystic involvement (● Fig. 1). The left ductal system was reported to be normal. After initial stabilization, the child underwent a right hepatectomy with a left hepaticojejunostomy. The postoperative period was uneventful, and at 8 years' follow-up the child is free of symptoms. Histopathology of the abscess wall showed that the other right duct cysts were free of any atypia.

Case 2

An 11-year-old boy presented with the classical triad of pain, jaundice and right hypochondrial mass. He had a history of 3 attacks of right upper quadrant pain in the last 2 years not associated with jaundice and was treated with analgesics and antacids by a local practitioner. Investigations revealed type IV-A choledochal cyst with selective left lobar involvement of the liver (● Fig. 2). The right duct system was essentially normal.

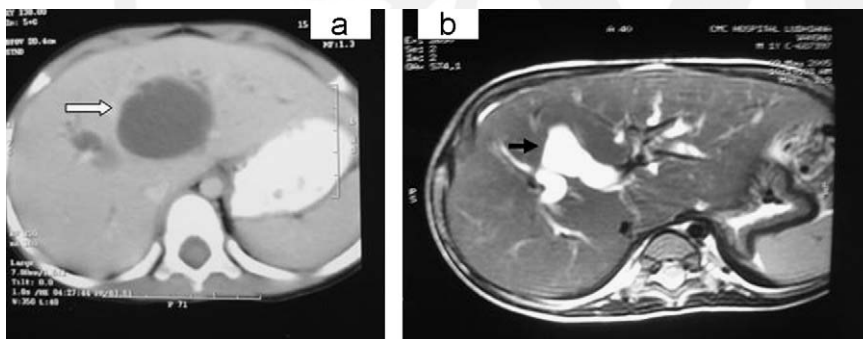


Fig. 1 CT (a) and MRI (b) scan showing a right lobar abscess and right ductal cysts with a normal left duct system.

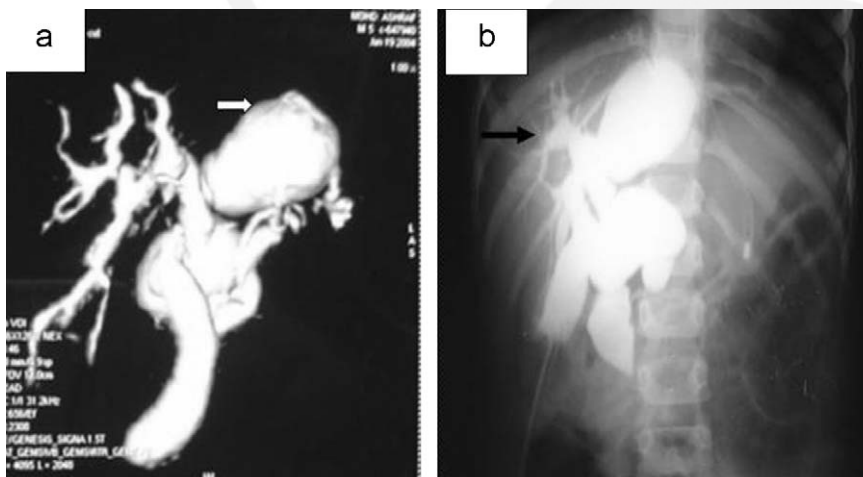


Fig. 2 MRCP (a) and intraoperative cholangiogram (b) showing Type IV-A cysts localized to the left ductal system (block arrow). The right ductal system is mildly dilated (arrow).

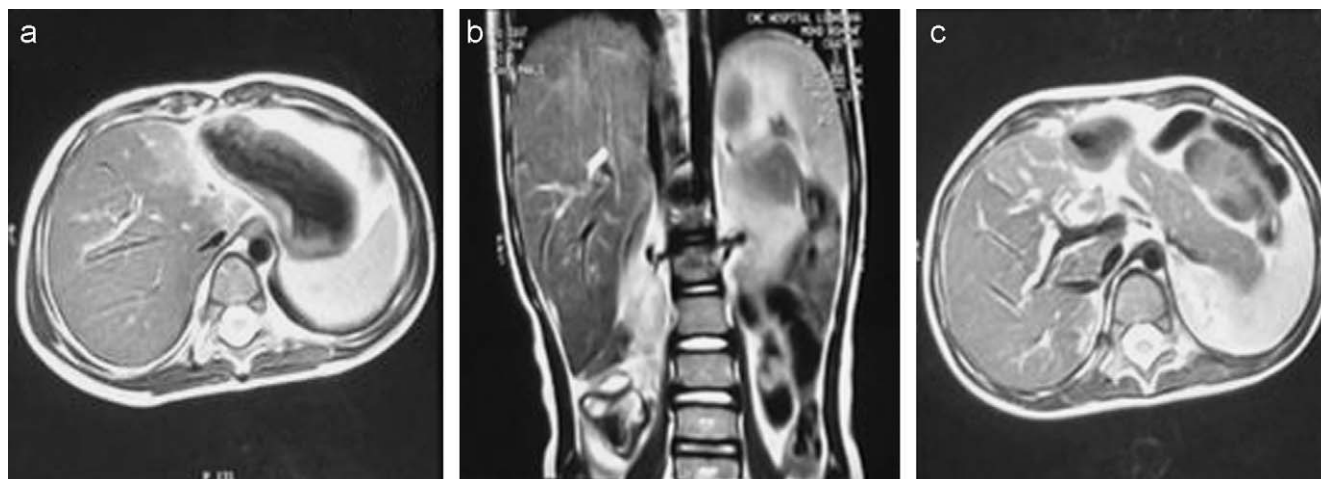


Fig. 3 Postoperative follow-up MRI showing hypertrophied right hepatic lobe with normal biliary ductal system.

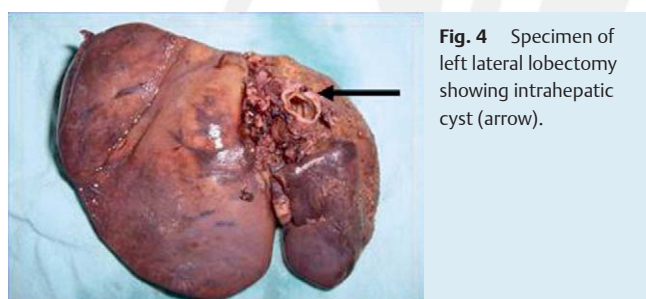


Fig. 4 Specimen of left lateral lobectomy showing intrahepatic cyst (arrow).

He underwent excision of an extrahepatic cyst, a left hepatectomy and a Roux-en-Y right hepaticojejunostomy. His postoperative recovery was unremarkable. Follow-up studies showed a normal right lobe ductal system (● **Fig. 3**) and the boy has remained completely free of clinical symptoms at 71/2 years' follow-up. Histopathology of the cysts showed them to be free from atypical/ dysplastic changes (● **Fig. 4**).

Case 3

A 5-year-old girl presented with fever, pain in the right hypochondrium and jaundice for 1 month's duration. Investigations revealed a huge extrahepatic cyst and a cystic affection of the confluence and left duct system extending and occupying most of the left lobe parenchyma (measuring 5×6 cm² in dimension). Mild back pressure changes were noticed on the right side.

The girl was managed by left hepatectomy, excision of an extrahepatic cyst including the confluence and Roux-en-Y right hepaticojejunostomy. Postoperative studies showed normalization of the right ductal dilatation within a period of one month, and the child has remained symptom-free at 5 years' follow-up. No abnormal finding (dysplasia/in situ carcinoma) in the cyst wall except for focal metaplasia in the extrahepatic cyst was reported on histopathology.

Case 4

A 10-year-old girl with thalassemia minor presented with fever, jaundice and pain in the right hypochondrium. Investigations revealed type IV-A choledochal cyst with intrahepatic cysts restricted to segment 2 and 3 of the left lobe. The right duct system was essentially normal. She was treated by excision of an extrahepatic cyst, a left lobectomy and Roux-en-Y hepaticojejunostomy. The postoperative period was marked by early cholan-

gitis treated by antibiotics. At 4 years' follow-up, she is a fully developed adolescent without any complaints. Histology of the cysts was normal.

Discussion



According to Todani et al., [13] congenital dilatation of the bile duct can appear anywhere in the biliary tree and is not confined to the choledochus. Accordingly, it has been suggested that these anomalies be named 'congenital bile duct cysts (dilatation)' and classified into six types. Types I-III correspond to Alonso-Lej et al.'s classification [1]. Type IV-A cysts are characterized as intrahepatic bile duct cysts and choledochal cysts. Type IV-B cysts consist of multiple cysts confined to the extrahepatic bile duct only, and in type V the cystic dilatations are confined to the intrahepatic bile ducts.

With the use of diagnostic modalities such as cholangiogram, ERCP, CT scan and MRCP, accurate anatomical diagnosis of these biliary duct cysts has led to a redefinition of the incidence of each type. Recent series [3,6,9,10,18] have reported an incidence of 33% to 45% for type IV-A cysts.

In our series, 10 out of 25 cases treated during a 5-year period were diagnosed as having type IV-A cysts (40%). Four of these 10 cases (40%) were in a localized form.

Excision of cysts and bilioenteric anastomosis

Management of biliary duct cysts has been refined over the last 3 decades. Internal drainage procedures for biliary cysts such as cholecystojejunostomy, choledochocystoduodenostomy, choledochocystojejunostomy and intrahepatic cystojejunostomy have been discarded by many. Intrahepatic cystojejunostomy is very occasionally performed as a palliative treatment for malignancy arising in the choledochal cyst to obtain free drainage of bile from intrahepatic cysts [6,9].

Excision of extrahepatic cysts and conventional biliary enteric anastomosis with the hepatic duct below the porta hepatis is the commonly practiced procedure. However, type IV-A cysts with intrahepatic involvement are often accompanied by a relatively narrow common hepatic duct (smaller than the size of the intrahepatic ductal dilatation). Consequently, conventional hepaticoenterostomy below the hilum often results in an anastomotic stricture with ascending cholangitis and intrahepatic cholelithiasis requiring additional procedures [9,14-16,19]. Recently,

wide hilar hepaticoenterostomy with or without ductoplasty or subtotal excision of the intrahepatic cyst and intrahepatic hepaticoenterostomy is considered the essential primary surgery for type IV-A cysts [1, 3, 9, 15]. Although the incidence of postoperative cholangitis is reduced with this modified approach, the risk of malignancy in the residual intrahepatic cysts remains the same.

Intrahepatic anatomical variations and biliary complications

Strong [12] has contested in his review that a bilioenteric anastomosis of adequate size does not eliminate the propensity to develop biliary complications at a later stage. Biliary sludge formation and intraductal lithiasis can result from the stagnation of bile in the saccular dilatation of the intrahepatic ducts. Chijiwa et al. [3] have confirmed the occurrence of frequent cholangitis with hepatolithiasis in the intrahepatic component of type IV-A cysts in the absence of an anastomotic stricture. In our series, one of the cases (case #1) underwent primary excision of an extrahepatic choledochal cyst and wide hilar anastomosis later complicated by a right hepatic abscess in the retained intrahepatic cyst. Another case (case #2) had left lobar involvement with an intrahepatic ductal stricture and saccular dilatations of the duct.

Type IV-A cysts are associated with a primary ductal stricture near the hilum, confluence or umbilical fissure requiring hilar ductoplasty and a large hepaticoenterostomy [14, 16, 19]. Although reported in up to 33% of type IV-A cysts, it is often overlooked because of an unsatisfactory cholangiogram. A membranous stricture at the umbilicus can be excised by ductal dissection from the hilum to the umbilicus with creation of a long cholangioenterostomy below the level of the porta and an intrahepatic cystoenterostomy at the hilum.

Biliary malignancy

Strong [12] and Benhidjeb et al. [2] have proposed that stasis, chronic inflammation and hepatolithiasis are associated with biliary malignancy. The biliary epithelium in the retained intrahepatic cysts is somewhat unstable and, with an appropriate trigger, is prone to malignant transformation. The mutagenicity of bile acids such as deoxycholate and lithocholate has been implicated.

The incidence of cholangiocarcinoma is reported to be up to 6.9% of type IV-A cysts after hepaticoenterostomy in the retained epithelium of the intrahepatic cysts [4, 5, 8, 15]. It is possible that cholangiocarcinoma in the retained intrahepatic cysts has been underreported. In situ carcinoma changes are evident even at 10 years of age and gross malignancy has been reported as appearing up to 25 years following initial surgery. Thus, young children will have a life span of many decades during which they carry this risk of malignant transformation.

The aim of surgery should be complete cyst excision, although this is achieved in only about 15-20% of cases with type IV-A cysts and Caroli's disease with unilobar involvement by a partial hepatectomy. In cases of bilobar cysts, dilatation of any intrahepatic stricture should be performed at the time of resection of any intrahepatic cyst and of hepatojejunostomy. If severe disease predominates in one area with bilobar disease, a partial hepatectomy should be performed. In the event of development of cirrhosis and portal hypertension or when recurrent and persistent hepatolithiasis or cholangitis complicate initial surgery

(resulting, for example, in hepatic abscess, rupture and malignancy), total hepatectomy and liver transplantation is indicated [3, 6, 10, 12-15, 18].

In a series of 17 patients who underwent resection of IV-A cysts, the only patient with a long-term survival (86 months) was a patient who had undergone a right hepatectomy for predominantly right-sided intrahepatic cysts [4]. It is clear that there are patients who may already have cancers in the intrahepatic cysts or who will develop cancer and who could benefit from a hepatectomy. Extensive involvement of the liver in persons with type IV-A or Caroli's disease will require a hepatectomy and transplantation as the ultimate curative procedure. Partial hepatectomy in the scenario of type IV-A cysts is a major decision, used sparingly by surgeons and only for intrahepatic abscess (due to stricture) as the risk of malignancy persists.

Unilobar variant of type IV-A cyst and hepatectomy

We have encountered a unique anatomical variation of type IV-A biliary duct cysts in 40% out of our total of 10 patients with type IV-A disease, in which the intrahepatic component was found to be localized to the left duct in three cases and to the right duct in one case. We achieved complete removal of the intrahepatic and extrahepatic cysts by left hepatectomy in 2, left lobectomy in 1 and right hepatectomy in one case. After 4-8 years of follow-up all the children remain free of symptoms and show no radiological changes in the biliary tree. Although theoretically the risk of malignancy in the biliary tree has been eliminated, long-term follow-up has been put in place to observe any complications. Recently, some authors [7, 11] have recommended more aggressive approaches such as excision of asymptomatic intrahepatic cysts, as they are of the opinion that secondary cyst excision may be more complicated and advanced malignant degeneration might be encountered during the secondary procedure. However, we are of the opinion that a judicious balance needs to be maintained between the decision regarding the extent of intrahepatic cyst excision and the surgical morbidity arising from the excision. In our first case we had to perform a secondary resection due to an abscess complicating the retained cyst. In three other cases the cysts occupied almost the entire segments subjected to resection and there was no significant morbidity associated with the procedures. This extra step during management has offered a clear advantage to our patients in the follow-up period.

Conclusion

Extrahepatic cyst excision accompanied by a hepatectomy is safe and technically easy in children with type IV-A choledochal cysts, including those who develop secondary intrahepatic complications (abscess, hepatolithiasis, cholangitis) following primary surgery, and offers a curative status for the unilobar variant with regard to future biliary complications and malignancy. It is imperative that surgeon must weigh the risk of cancer from residual intrahepatic cysts, as well as the risk of cholangitis, hepatolithiasis and abscess from deep intrahepatic strictures and morbidity from total cyst excision involving partial or extended hepatectomy. Total cyst excision is the ideal treatment if it can be accomplished without too much morbidity. Patient age, co-morbidities, anatomy and the surgeon's judgment should dictate the management.

Conflict of interest: None.

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