

Choledochal cysts in adults and their complications

HDE Atkinson, CP Fischer, CHC de Jong, KK Madhavan, RW Parks and OJ Garden

Department of Clinical and Surgical Sciences (Surgery), University of Edinburgh, Royal Infirmary of Edinburgh, Edinburgh, Scotland

Background

Despite refinements in the management of choledochal cysts in children, an increasing number of patients present with ongoing symptoms in adult life. The aim of this study was to review the management of adult patients with choledochal cysts in a tertiary referral centre.

Method

A retrospective review was carried out of all adult patients presenting with choledochal cysts to this department between 1992 and 2000. Patient records were reviewed and detailed analyses were made of the clinical presentation, radiological and biochemical findings, anatomical anomalies, management, complications and outcomes.

Results

Of 16 patients (12 women and 4 men; median age 23 years), 8 had undergone previous upper gastrointestinal operations before referral, including 5 who had had previous cyst drainage procedures. All patients underwent elective complete cyst

excision with Roux-en-Y hepaticojejunostomy. There were no operative deaths and there was a low early postoperative morbidity rate (25%). There was no evidence of biliary malignancy in any cyst. During a median postoperative follow-up of 44 months, five patients (31%) continued to experience cholangitis and two of these required additional revisional procedures, but are now symptom-free.

Conclusion

Patients with choledochal cysts should be referred to specialised tertiary surgical units. Total choledochal cyst excision with Roux-en-Y hepaticojejunostomy is the treatment of choice. Patients with previous inadequate cyst excisional procedures should undergo revisional surgery, to reduce recurrent symptoms and the risk of developing cholangiocarcinoma.

Keywords

choledochal cyst, cholangitis, hepaticojejunostomy, cholangiocarcinoma, cystduodenostomy

Introduction

Choledochal cysts are rare cystic transformations of the biliary tree that are increasingly diagnosed in adult patients [1–4]. These congenital lesions have a female:male preponderance of 4:1, with an incidence in the West of between 1 in 50 000 and 1 in 200 000 live births [3]; they are much more frequent in Asia [5, 6]. Nearly 25% of choledochal cysts are detected in the first year of life and 60% in the first decade, but 20% are diagnosed after the age of 20 years [1, 7].

Adults with cysts are especially predisposed to the development of cholangiocarcinoma, with the cancer incidence increasing from 0.7% in the first decade of life to >14% after the age of 20 years [8–10]. Several reports have documented long-term complications following the paediatric management of choledochal cysts using cystenterostomy without total cyst excision [8, 11, 12]. Many centres now see an increasing number of adult patients who have either had previous operative procedures (without cyst excision) or experience ongoing

complications related to a cyst in situ [7, 13–16]; up to 80% experience recurrent cholangitis and pancreatitis.

Because of the risk of malignant change in choledochal cysts, the ideal treatment is excision of the entire dilated biliary tree with reconstruction using a Roux-en-Y hepaticojejunostomy [3, 9, 10, 17]. We present a contemporary cohort of adult patients with choledochal cysts, which emphasises the complexity and diversity of this disorder in adults.

Methods

In a retrospective review of all adults presenting with choledochal cysts to the Royal Infirmary of Edinburgh, Scotland, between 1992 and 2000, patients were identified by means of the Lothian Surgical Audit and their medical and operative records were reviewed. Detailed analyses were made of the clinical presentation, radiological and biochemical findings, anatomical anomalies, management, complications and outcomes.

Investigation of the patients was dictated by their

Correspondence to: RW Parks, Department of Clinical & Surgical Sciences (Surgery), Royal Infirmary of Edinburgh, Edinburgh, EH3 9YW, UK (e-mail: R.W.Parks@ed.ac.uk)

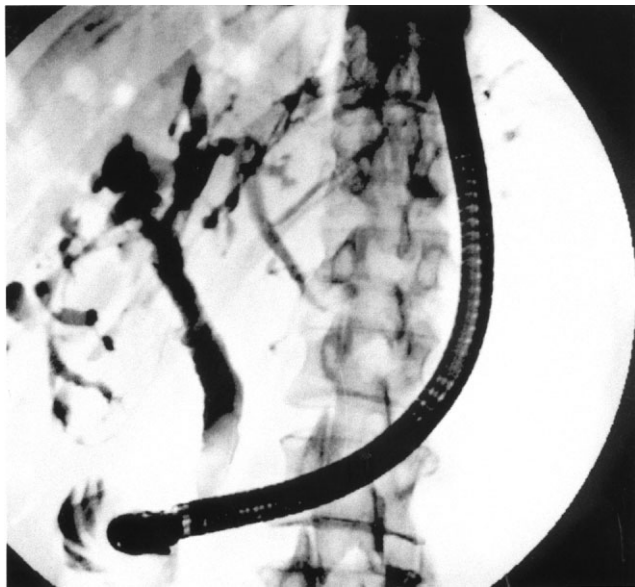


Figure 1. ERCP demonstrating extrahepatic and intrahepatic biliary duct dilatation in a type IV choledochal cyst.

presentation, but generally involved assessment of the biliary tree by endoscopic retrograde cholangio-pancreatography (ERCP) (Figure 1) and computed tomography (CT) scanning (Figure 2).

Patients with choledochal cysts in situ underwent total cyst excision with reconstruction of the biliary tract by means of a Roux-en-Y hepaticojejunostomy. Patients with Caroli's disease were excluded from the series.

Results

Sixteen patients (12 women and 4 men) were identified, with a median age of 23 years (range 18–49 years). According to the modified Todani classification, 13 patients had type I cysts and 3 had type IVa cysts (Figure 3).

Eight of the 16 patients had undergone operations before referral to this institution (Table 1). One of these patients had presented previously to a local hospital as an adult with severe abdominal pain and underwent laparotomy without notable findings. In retrospect, he probably had cholangitis and an infected choledochal cyst. A second patient had presented with a history of recurrent idiopathic pancreatitis and had undergone transduodenal sphincteroplasty and cholecystectomy, but the relevance of her choledochal cyst was not appreciated at that time. One patient underwent cholecystectomy for recurrent upper abdominal pain, again without diagnosis or recognition of the choledochal

cyst. The remaining five patients underwent cystoduodenostomy for known choledochal cysts in childhood. The cyst (or part of the cyst) remained in situ in all patients who had undergone previous operation.

All patients presented with symptoms of recurrent abdominal pain. Other symptoms included recurrent nausea in 13, vomiting in 8, dyspepsia in 4 and weight loss in 3. At referral, 10 patients had a history of at least one episode of cholangitis and 5 had previous pancreatitis. Median duration of symptoms before referral was 10 months (range 1 week to 6 years). Abnormal liver function tests were recorded in 69% of patients at the time of diagnosis. These levels did not remain consistently abnormal, and all patients had normal laboratory tests at the time of operation.

All patients underwent elective definitive surgery in this unit, comprising complete cyst excision and Roux-en-Y hepaticojejunostomy. In all but one case, the biliary-enteric anastomosis was fashioned at the biliary confluence using interrupted 4/0 PDS sutures. One patient, who had had dense adhesions and scarring following a previous procedure, underwent a single anastomosis to include the left hepatic duct. No patient required separate anastomoses to the right and left hepatic ducts, and no internal stents or access loops were employed in any of the patients. The 12 patients with intact gallbladders also underwent cholecystectomy.

A number of anomalies were identified during investigation or at operation (Table 2), including the presence of an anomalous pancreatobiliary junction



Figure 2. CT scan demonstrating intrahepatic biliary duct dilatation in a type IV choledochal cyst.

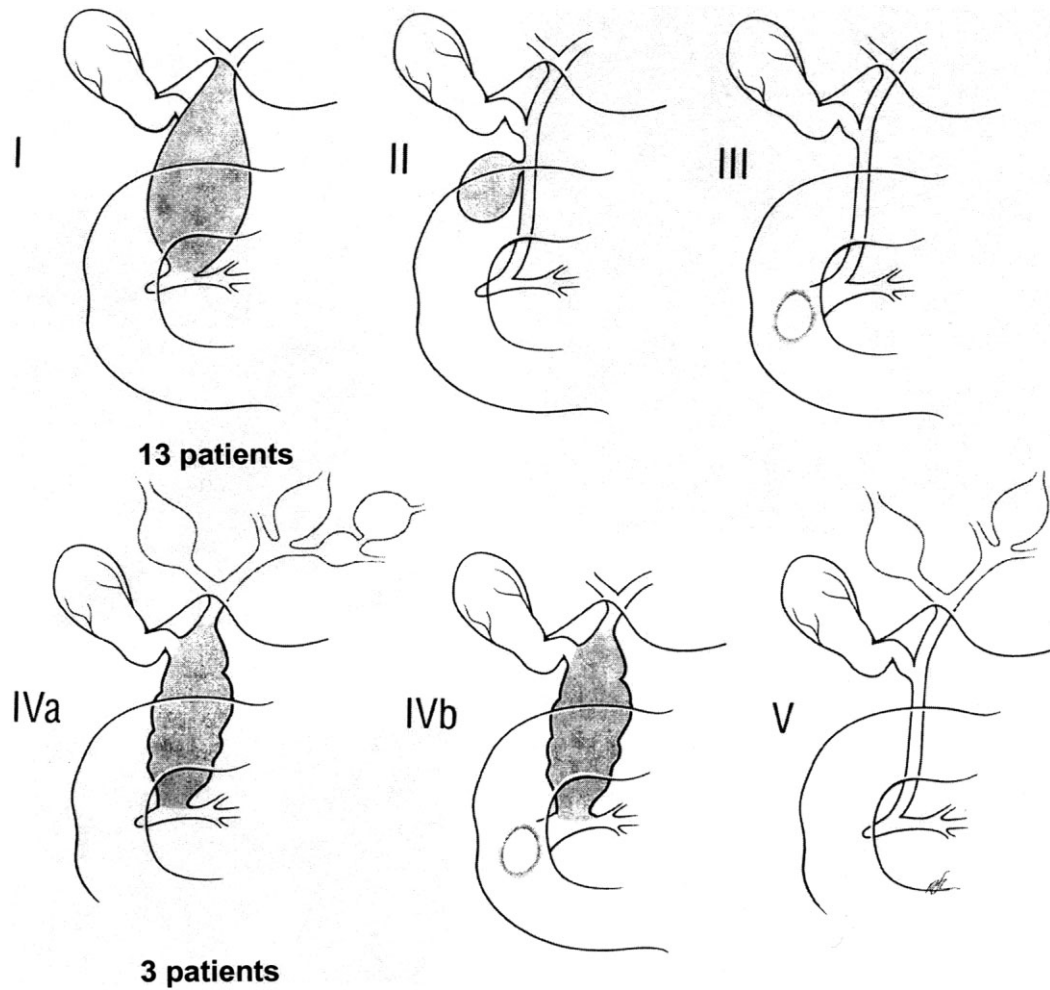


Figure 3. The modified Todani classification, and distribution of choledochal cysts in this series.

Table I. Patient characteristics, previous operations, follow-up and complications

Age (years)	Sex	Todani classification	Previous operation	Length of follow-up (months)	Complications	Cholangitis	Re-operation
30	M	Type I		111		+	
49	F	Type I	Partial cyst excision and cystduodenostomy	101			
20	M	Type I		51	Subphrenic abscess		
19	F	Type I	Partial cyst excision and cystduodenostomy	51		+	+
23	F	Type I		44			
36	F	Type I	Cholecystectomy	40			
32	F	Type I	Cystduodenostomy and cholecystectomy	37	Mild pancreatitis		
18	F	Type I		35	Bile leak		
23	F	Type I	Cystduodenostomy and cholecystectomy	33		+	+
31	M	Type I	Negative exploratory laparotomy for right upper quadrant pain	29	Bile leak		
19	F	Type I		27			
31	F	Type I	Transduodenal sphincteroplasty and cholecystectomy	23			
21	F	Type I		21			
18	M	Type IVa	Cystduodenostomy	105		+	
31	F	Type IVa		90			
31	F	Type IVa		51		+	

(APBJ) (10 patients), an aberrant hepatic artery (4 patients), primary ductal stricture (2 patients), a prominent accessory papilla (1 patient) and an accessory gallbladder (1 patient). In addition, seven patients had cholelithiasis, six patients had stones within the choledochal cyst and two patients had intrahepatic stones. Histological assessment confirmed choledochal cyst in all cases with no evidence of malignancy.

There were no operative deaths, while 4 of the 16 patients (25%) had minor postoperative complications (Table 1). Two patients developed bile leaks and one patient had an episode of mild pancreatitis. These three complications settled spontaneously without the need for further intervention. One patient developed a subphrenic collection, which was drained percutaneously. The median postoperative length of hospital stay was 8 days (range 6–12 days).

During a median postoperative follow-up of 44 months (range 21–111 months), five patients had at least one further episode of cholangitis (Table 1). Three of these patients had type I cysts (two with previous cystduodenostomy) and two had type IVa cysts (one with previous cystduodenostomy). Two of the symptomatic patients have required revisional surgery. The first patient had a type I cyst treated previously with partial cyst excision and cystduodenostomy 12 years before definitive surgery in this unit. She subsequently developed intrahepatic ductal calculi and required anastomotic revision and stone extraction from the left duct system. The second patient also had a type I cyst and had undergone previous cystduodenostomy as a child. She developed an anastomotic stricture and required revisional surgery. Both patients remain symptom-free at 33 and 51 months after re-operation.

Table 2. Hepatobiliary pathology associated with the choledochal cysts in this series

Pathology	Number	Percentage
Anomalous pancreato-biliary junction	10	63
Primary ductal stricture	2	13
Secondary ductal stricture	5	31
Aberrant hepatic artery	4	25
Prominent accessory papilla	1	6
Accessory gallbladder	1	6
Cholelithiasis	7	44
Cystolithiasis	6	38
Intrahepatic stones	2	13

Discussion

We present the management of a series of adult patients with choledochal cysts and symptoms related to the presence of the cyst in situ. Their presentations did not follow any classical patterns. Many had episodic and non-specific symptoms, but all experienced abdominal pain and most had episodes of cholangitis. Although 69% had abnormal liver function tests, none of these were consistently abnormal. This fact may explain why the diagnosis was often delayed, and inappropriate upper gastrointestinal surgery (with incorrect diagnoses) was performed in three of the eight patients who had been operated elsewhere. Five patients had previously undergone inadequate procedures for their choledochal cysts in the form of cystduodenostomy and had recurrent symptoms related to the cyst remnant. The literature reports that up to 80% of such patients will have recurrent symptoms [15, 16] and that up to 69% will subsequently require revisional surgery [18–20].

Although early postoperative morbidity was low, five patients (31%) developed recurrent cholangitis and two (13%) have required additional revisional procedures at our institution. This finding emphasises the difficulty in managing adult patients with this condition and the dangers of exposing the patient to the risks of secondary sclerosing cholangitis following an inappropriate operation.

Yamataka and co-workers [21] reported a 23% incidence of postoperative cholangitis after primary definitive cyst excision of type I cysts, while other authors have reported the incidence to range up to 40% in adults [4, 15, 22]. This figure is even higher for type IVa cysts, ranging from 31% to 44% [3, 22, 23]. Specific causes of recurrent cholangitis after definitive cyst excision have been postulated by Todani and colleagues [22]. In this series, only one of the eight patients with type I cysts who had undergone a primary definitive procedure developed recurrent cholangitis, whereas two further patients with type I cysts suffered recurrent cholangitis after cystduodenostomy. Of the two patients with type IVa cysts experiencing ongoing problems, one had undergone previous surgery. These overall results compare favourably to the literature, giving a rate of 18% for recurrent cholangitis if the primary definitive procedure is undertaken in a specialist unit.

There are limited data in the literature with regard to

recurrent symptomatology following revisional surgery in those who have previously had cyst drainage procedures. The fact that three of the six revision patients had recurrent cholangitis following a definitive procedure suggests that an inadequate previous operation may compromise long-term results.

In this series, 40% of postoperative cholangitis was caused by the development of an anastomotic stricture, despite this being fashioned at the biliary confluence in accordance with previous recommendations [22, 24]. These complications appear more frequently in patients who have undergone previous cyst drainage procedures, especially those patients with type IVa cysts [21, 25, 26]. The two patients who required re-operation and anastomotic revision in this series had both had cystoduodenostomies performed in childhood. The extensive fibrosis and shortening of the hepatoduodenal ligament noted in both cases has been suggested by other authors to be an independent risk factor for postoperative stenosis and cholangitis [21, 22]. Two further patients with recurrent cholangitis had type IVa cysts, which are known to promote biliary stasis and stone formation due to intrahepatic cyst disease, which may not be adequately drained by hepaticojejunostomy [21, 25–27]. Intrahepatic calculi were identified in both these patients.

The aetiology of choledochal cysts remains somewhat elusive. One aetiological factor may be the reflux of pancreatic juice into the common bile duct (CBD), possibly due to the presence of an anomalous pancreatobiliary junction (APBJ), which is frequently associated with choledochal cysts [28–30]. It is hypothesised that this anomaly leads to weakening of the duct wall and causes inflammatory changes within the endothelium [31], although an experimental model failed to confirm this finding [32]. This theory also seems to be refuted by the case report of a choledochal cyst diagnosed at 15 weeks in utero, i.e. before the exocrine function of the pancreas had even begun [33]. However, the strong association of an APBJ with choledochal cysts cannot be ignored [34]; 63% of our patients had APBJ either diagnosed by preoperative cholangiography or intraoperatively. Reports of the observed incidence of this abnormality vary from as low as 44% in adult patients [35] to nearly 100% in some paediatric populations [36]. The high association with APBJ in children emphasises the potential association, but the lesser frequency with which it is observed in adults suggests other aetiological

factors in this patient group. The other anomalies identified at operation or during investigation were also strongly associated with choledochal cyst [14].

We observed no malignancy in this group of patients, yet the incidence in the Asian literature ranges from 8% to 19% [11, 12, 24], with similar figures in some Western series [3, 7]. The median age of our patients was 23 years (mean 27 years), which is lower than the mean age of approximately 45 years in other reports [11, 37] and may explain why cholangiocarcinoma was not encountered.

There are many reports of malignancy in patients previously treated by cyst drainage procedures [9, 11, 17], and there is growing evidence that these patients may be at an increased risk of developing malignancy [3, 12, 20, 28], presenting 15 years earlier than those who have had no procedure [37]. This phenomenon may be due to reflux of gut bacteria and enzymes into the bile ducts via the enteric anastomosis. These reports emphasise the importance of avoiding cyst drainage procedures and of undertaking complete choledochal cyst excision. Revisional surgery should include definitive cyst excision with a Roux-en-Y hepaticojejunostomy to reduce the likelihood of malignancy. Interestingly, the risk of malignancy in those patients with type IVa cysts is negligible following complete extrahepatic ductal excision with Roux-en-Y hepaticojejunostomy, even though the intrahepatic disease remains in situ, while this figure approaches 45% in patients if no treatment is undertaken [38].

The management of choledochal cysts in adult surgical units is being increasingly reported. The high rate of cholangitis and need for revisional surgery that we and others have reported emphasises that these cases are best managed in specialised tertiary surgical units [22, 23, 39].

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