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Surgical Strategy for Cystic Diseases of the Liver in a Western Hepatobiliary Center

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Abstract. The aim of this study was to define the indications and evaluate the results of various management options in patients with cystic liver disease. Between 1992 and 1999 we managed 60 consecutive patients with cystic liver disease. Diagnoses included a simple cyst (solitary 12, multiple 10), adult polycystic liver disease (APLD 17), Caroli's disease (8), hydatid cysts (4), and neoplastic cysts (9). Half of the patients with simple cysts had mild or no symptoms and required no treatment. Percutaneous drainage in eight patients (simple cyst 4, APLD 4) was followed by symptomatic recurrence in three. Laparoscopic deroofing in three patients (multiple simple cysts 2, APLD 1) was followed by symptomatic enlargement of the remaining cysts that required further intervention (laparoscopic deroofing 2, transplantation 1). Laparoscopic hepatectomy was successful in three patients with solitary simple cysts. Of 18 patients who underwent open hepatic resection (neoplastic 8, Caroli's 4, simple cysts 3, hydatid cysts 2, APLD 1), 2 patients with Caroli's disease required liver transplantation for disease progression. Nine patients (Caroli's 5, APLD 4) underwent liver transplantation, and three had a concomitant renal transplant. Seven patients developed complications, and three died (5%). Cholangiocarcinoma developed in three patients with bilateral Caroli's disease, and all died. Radiologic treatment has a limited role in the management of patients with simple cysts or APLD. Laparoscopic deroofing of simple cysts may have to be repeated, whereas resection minimizes cyst recurrence. Unilobar Caroli's disease may be resected, whereas bilateral disease requires early liver transplantation owing to the high risk of malignancy. Transplantation is a reserved option in patients with extensive APLD.

Liver cysts are not uncommon, occurring in approximately 0.1% of the population [1]. Although some may attain large sizes and become symptomatic, others remain asymptomatic and present frequently as an incidental finding on ultrasonography [2] or computed tomography (CT). Cystic lesions of the liver represent a wide spectrum of diseases ranging from simple benign cysts, to potentially malignant biliary cysts, to de novo malignant cystic tumors [3].

We present our experience with the management of cystic diseases of the liver in a Western tertiary referral center serving a population where hydatid disease is extremely uncommon. Our limited experience with the treatment of hydatid disease is only briefly mentioned, and the issue is not discussed further; the reader is referred to some of the recent articles on the minimally invasive management of this condition [4–6]. The roles of various management options (observation or discharge, radiologic drainage, laparoscopic deroofing or liver resection, liver transplantation) are discussed. The potential for malignant transformation in patients with Caroli's disease is emphasized. A management algorithm for cystic liver diseases is proposed.

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Methods

Between July 1992 and April 1999 a series of 60 consecutive patients were referred to our Centre of Hepatobiliary and Transplantation Surgery for the management of cystic liver disease. The medical records of the patients were reviewed; and data including demographic features, symptoms, biochemical and radiologic investigations, and management were collected.

Assessment

The aims of the assessment were fourfold: evaluate the severity of symptoms; establish the diagnosis; assess the impact of disease on liver function; and establish fitness for surgery if it is indicated. The radiologic studies employed included (1) routine ultrasonography to determine the number, size, and nature of the cysts and the status of the biliary tree, and (2) magnetic resonance imaging (MRI) or computed tomography (CT) to establish the nature of the cysts suspected to be malignant and to assess their resectability. Serologic studies for hydatid disease and tumor markers [carcinoembryonic antigen (CEA), CA 19-9] were undertaken when indicated. Biochemical assessments of liver function were done routinely, as was serology for hepatitis A, B, and C. Percutaneous sampling of the cyst fluid for cytology was avoided. Cystic dilatation of the intrahepatic bile ducts (Caroli's disease [7]) was demonstrated by endoscopic retrograde cholangiopancreatography (ERCP) or by a hepatic iminodiacetic acid (HIDA) scan.

Classification

Cystic diseases of the liver were classified as simple (solitary or multiple), polycystic, neoplastic (cystadenoma, cystadenocarcinoma, cystic sarcoma), or parasitic (hydatid disease).

Management

Conservative management with clinical and radiologic follow-up was adopted in asymptomatic patients with solitary or multiple cysts and in those with adult polycystic liver disease (APLD). Patients with symptomatic solitary or multiple cysts and those with large cysts related to APLD were submitted for percutaneous drainage or laparoscopic deroofing. The laparoscopic approach was favored in younger, fitter patients, in patients with dominantly large cyst(s), and after failed (recurrence) radiologic drainage. Cysts located in the anterior liver segments are best suited for laparoscopic treatment. The radiologic approach was reserved for older, frail subjects, multiple but nondominant cysts, cysts predominantly situated in the posterior aspect of the liver where access at laparoscopy may be limited, when doubt existed as to the relation of cysts to symptoms, and for patients who refused surgery. Liver resection was employed for symptomatic localized disease that involved most of a lobe or segments and when other, less invasive therapies had failed or were contraindicated.

Patients with severely symptomatic APLD in whom cyst(s) deroofing or liver resection had failed or who were considered unsuitable underwent orthotopic liver transplantation; simultaneous kidney transplantation was employed in patients with concomitant end-stage renal failure secondary to adult polycystic renal disease. Other indications for surgery included parasitic (hydatid) cysts and suspected or confirmed neoplastic cysts. Patients with hydatid disease of the liver were given albendazole (800 mg daily) orally for 28 days prior to surgery. Monolobar or segmental symptomatic Caroli's disease was treated by liver resection, whereas bilateral and more extensive disease complicated by recurrent cholangitis, cirrhosis, or both was managed by liver transplantation with excision of the extrahepatic biliary tree and Roux-en-Y hepaticojejunostomy.

Results

There were 14 male and 46 female patients, with a mean age of 54 years (range 23–85 years). The modes of presentation were abdominal pain in 45 patients (75%), abdominal distension in 28 patients (47%), vomiting in 10 patients (17%), and recurrent cholangitis in 7 patients (12%) with Caroli's disease (associated with gastrointestinal bleeding in 1 patient). Liver cyst(s) were detected incidentally during ultrasound examination of the abdomen in 14 patients (23%).

The etiologies of liver cysts in the 60 patients are summarized in Table 1. The management of these patients is summarized in Table 2.

Table 1. Etiology of cystic liver disease in 60 patients.

Etiology	No. of patients
Solitary simple cyst	12 (20.0%)
Multiple simple cysts	10 (16.7%)
Polycystic disease	17 (28.3%)
Caroli's disease	8 (13.3%)
Hydatid cyst	4 (6.7%)
Cystadenoma	6 (10.0%)
Cystadenocarcinoma	2 (3.3%)
Malignant hepatoblastoma	1 (1.7%)

Simple Cysts

Simple hepatic cysts were detected in 22 of 60 patients (36.7%); they were solitary (Fig. 1) in 12 patients and multiple (Fig. 2) in 10. There were 20 women and 2 men, with a mean age of 62.5 years (range 40–85 years). Fifteen patients (68%) had symptomatic disease, with abdominal pain (68%) and distension (50%) being the commonest presenting symptoms. Abdominal pain was associated with bleeding into the cyst in one patient. The disease was asymptomatic and incidental in seven patients (32%). The cysts varied widely in size; the maximum diameter of the solitary cysts was 20 cm, and that of multiple cysts was 17 cm.

Conservative treatment was adopted in 11 patients (solitary cyst 6, multiple cysts 5) with mild ($n = 4$) or no ($n = 7$) symptoms. None of these patients required intervention, and most were discharged after a median follow-up of 28 months. Radiologic drainage in four patients with symptomatic cysts (solitary cyst 1, multiple cysts 3) was followed by cyst recurrence in two patients, one of whom underwent laparoscopic deroofing; the other remained asymptomatic.

Surgical intervention was required in eight patients (laparoscopic 5, open 3). Laparoscopic management included deroofing the cyst(s) in two patients with multicystic disease and hepatic resection in three patients with solitary cysts (left lateral sectionectomy 2, left hemihepatectomy 1). Three symptomatic patients who were suspected to have cystadenoma but were subsequently found to have large simple cysts underwent open hepatic resection (left trisectionectomy, left lateral sectionectomy, segmentectomy). One patient who underwent an emergency left hepatic lateral sectionectomy for a massive hemorrhage into the cyst with intraperitoneal rupture died 8 days postoperatively from aspiration pneumonia and cardiopulmonary failure.

Follow-up was available for 27 to 80 months following surgical treatment (median 45 months). Other small (< 1.5 cm in diameter) hepatic cysts, one to three in number, were detected on ultrasonography in two patients following laparoscopic hepatic resection for a solitary simple cyst. They remained asymptomatic and required no further intervention. Further enlargement of some of the residual cysts in two patients with multicystic disease with recurrence of abdominal pain 6 and 9 months following laparoscopic deroofing necessitated further laparoscopic deroofing, which led to satisfactory symptomatic relief.

Adult Polycystic Liver Disease

Adult polycystic liver disease (Fig. 3) was observed in 17 of 60 patients (28.3%). There were 13 women and four men, with a mean age of 50 years (range 31–72 years). Renal cysts coexisted in 8 patients (47%). Abdominal pain, with or without abdominal distension, was the presenting symptom in 11 patients (65%); and in the remaining 6 patients (35%) the cysts were an incidental finding on ultrasonography.

No intervention was required in 5 of 17 patients with minimal or no symptoms and preserved liver and renal functions. Percutane-

Table 2. Summary of the management of 60 patients with cystic diseases of the liver.

Parameter	Solitary simple cyst	Multiple simple cysts	Polycystic liver disease	Caroli's disease	Hydatid cyst	Cystic neoplasms
No. of patients	12	10	17	8	4	9
Management						
Conservative	6	5	5	1	1	1
Radiologic drainage	1	3[a]	4[b]	—	—	—
Pericystectomy	—	—	—	—	1	—
Laparoscopic deroofing	—	2	1[c]	—	—	—
Laparoscopic liver resection	3	—	—	—	—	—
Open liver resection	2	1	1	4[d]	2	8
Liver transplantation	—	—	2	4	—	—
Kidney transplantation	—	—	4	—	—	—
Liver and kidney transplantation	—	—	2	1	—	—

[a,b]Cyst recurrence after radiologic drainage in one patient with multiple simple cysts[a] was treated by laparoscopic deroofing and in another patient with adult polycystic liver disease[b] by partial hepatectomy.
[c]Laparoscopic deroofing was succeeded by deterioration in liver function that necessitated an orthotopic liver transplantation.
[d]Caroli's disease recurred in the remaining liver in two patients who subsequently underwent liver transplantation.

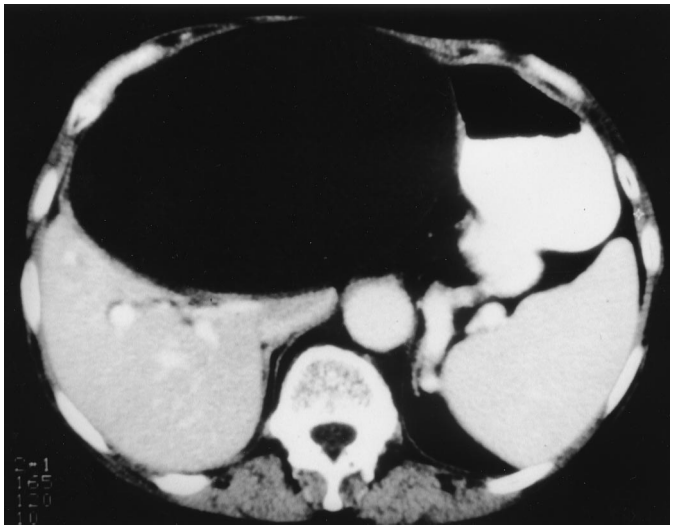

Fig. 1. Large solitary simple cyst treated by laparoscopic left hemihepatectomy.

ous drainage of symptomatic and extremely large cysts was carried out in four patients (one to three procedures per patient) with good symptomatic response in three. Following three percutaneous aspirations with instillation of tetracycline into the cysts, the fourth patient underwent right hemihepatectomy for a recurrent, large cyst (12 cm) with relative sparing of the left hemiliver. Laparoscopic fenestration in a 45-year-old patient with gross hepatic involvement and intractable abdominal pain conferred no symptomatic benefit and was succeeded by deterioration of liver function for which orthotopic liver transplantation was performed. In total, eight patients underwent organ transplantation (liver 2, liver and kidney 2, kidney 4). They included three of six patients who were initially managed expectantly but later required renal transplantation. Impaired renal function created complications in six of the eight patients with concomitant hepatic and renal cysts, two of whom underwent simultaneous liver and kidney transplantation; four others underwent renal transplantation.

A 49-year-old patient died during liver transplantation from cardiac failure. There were no other complications in this group of patients, and the 16 remaining patients are alive and asymptomatic at 20 to 88 months following surgical or radiologic intervention (or both) (median follow-up 36 months).

Caroli's Disease

Eight patients (13.3%) underwent surgery for Caroli's disease. There were six female and two male subjects, age 23 to 55 years (mean age 35 years). The disease was segmental in three patients and multilobular in five patients. Abdominal pain and recurrent episodes of cholangitis were the presenting symptoms. In addition, one patient had hemobilia with overt gastrointestinal bleeding. Four patients had previously undergone cholecystectomy and one to three biliary reconstruction procedures. Another patient had undergone kidney transplantation for polycystic renal disease during childhood 17 years earlier. All patients had evidence of cystic dilatation of the intrahepatic bile ducts that contained calculi, as seen on ERCP.

Liver resection was carried out in the three patients with segmental disease (left lateral sectionectomy 2, right posterior sectionectomy 1). The disease recurred in the remaining liver in two patients and presented with abdominal pain and repeated episodes of cholangitis that culminated in the development of secondary biliary cirrhosis 2 and 9 years, respectively, after surgery; both were treated by liver transplantation with Roux-en-Y hepaticojejunostomy. The explanted liver of the latter patient showed histologic evidence of cholangiocarcinoma, and the patient died 16 months after transplantation.

One patient with symptomatic bilobar disease underwent right hepatic trisectionectomy but developed portal vein thrombosis with secondary bleeding from esophageal varices a few months later. She received endoscopic sclerotherapy and was placed on the waiting list for liver transplantation but succumbed abroad a year after her surgery with hematemesis.

Four patients with bilobar disease and liver cirrhosis underwent orthotopic liver transplantation with a Roux-en-Y hepaticojejunostomy as the primary treatment modality; one patient required simultaneous kidney transplantation for end-stage renal failure associated with medullary sponge kidneys. The explanted liver of the latter patient showed multiple foci of cholangiocarcinoma, and the patient died 3 months after surgery. Peritoneal seedlings

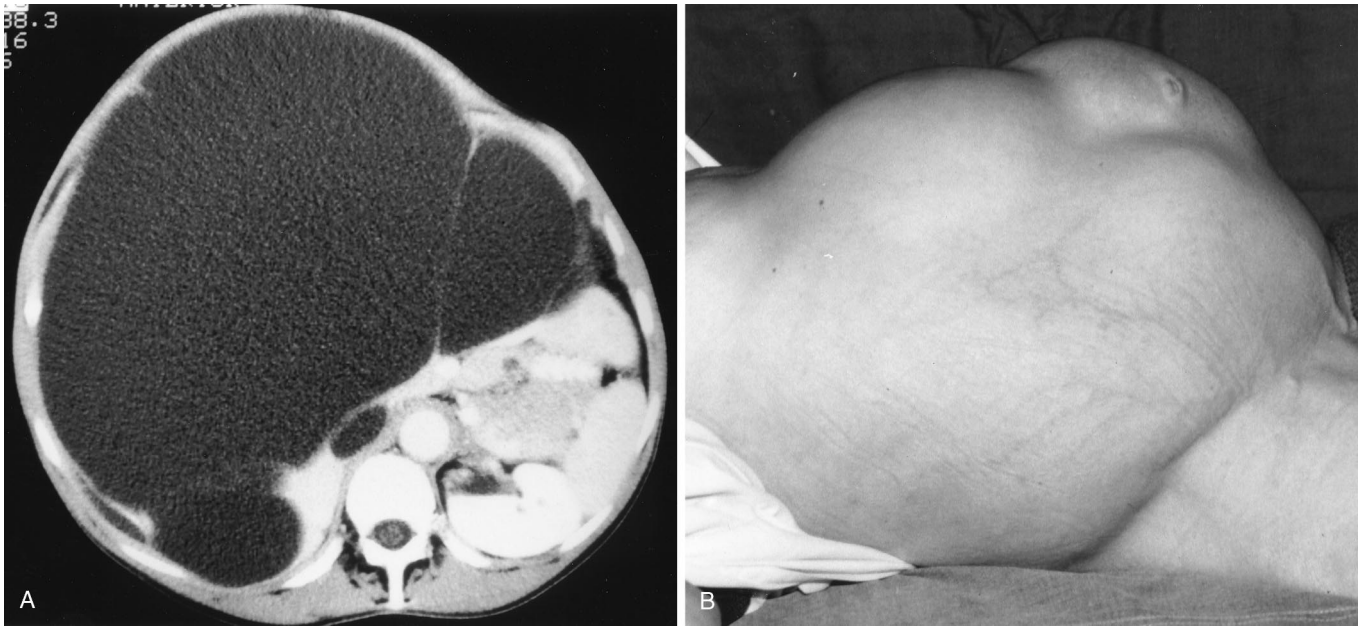

Fig. 2. Computed tomography (CT) reveals multiple simple cysts (**A**) associated with massive abdominal distension (**B**). They were treated by laparoscopic deroofing. Further enlargement of residual cysts necessitated another laparoscopic deroofing 9 months later.

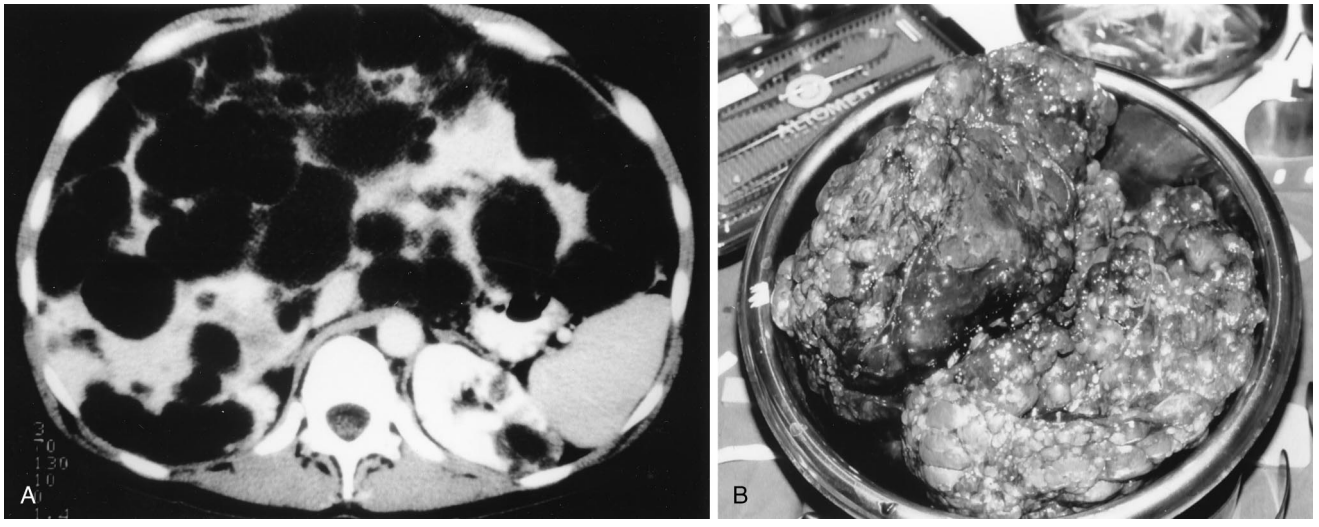

Fig. 3. A. CT appearance of polycystic liver disease associated with minor renal cystic disease. The patient required orthotopic liver transplantation. **B**. Explanted liver.

from a cholangiocarcinoma were detected at laparotomy with intention to liver transplantation in a fifth patient with diffuse Caroli's disease (and previous bile duct reconstruction) and led to the patient's demise 4 months later. Four patients remain alive at a median follow-up of 20 months (15–32 months).

Parasitic (Hydatid) Cysts

Hydatid cysts of the liver were uncommon (four patients, 6.7%), reflecting UK practice. Three patients were immigrants from Pakistan and Iraq, where hydatid disease is prevalent. There were two men and two women, with a mean age of 41 years (range 31–59 years). Right upper abdominal pain was the presenting symptom in three patients, and the cyst was found incidentally at abdominal ultrasonography for menorrhagia in one patient. There was one cyst in each of two patients, two cysts in one patient, and multiple cysts in one patient. Two patients underwent formal hepatic resection (left hemihepatectomy and right hemihepatectomy, respectively), and a third underwent partial cystectomy and omentoplasty. No complications developed. The fourth patient was asymptomatic and declined surgery. There has been no recurrence of cysts at a median follow-up of 18 months (range 12–42 months).

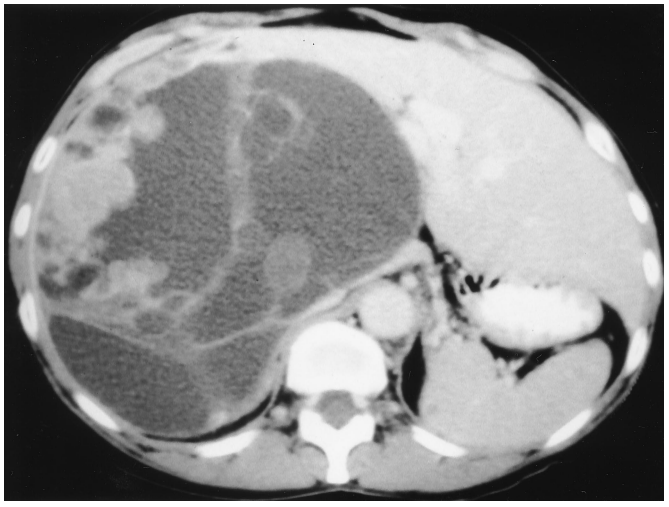

Fig. 4. CT appearance of a cystadenocarcinoma of liver that was aspirated several times over a 14-year period before referral. The patient underwent right hepatic trisectionectomy and remains disease-free at 6 years.

Neoplastic Cysts and Cystic Neoplasms

Eight patients with neoplastic cysts (cystadenoma 6, cystadenocarcinoma 2) and one patient with cystic neoplasm (embryonic sarcoma) were treated (Fig. 4). There were four men and five women, with a mean age of 50 years (range 23–73 years). Abdominal pain and distension were the presenting symptoms in all nine patients. Hemorrhage into a "cyst" resulted in the development of severe abdominal pain and swelling in the patient with an embryonic sarcoma. The cyst was an incidental finding on ultrasonography in one patient. The cysts were solitary in eight patients and multiple in one patient with metastatic cystadenocarcinoma. The sizes of the cysts ranged between 3 and 18 cm (median 12 cm).

The involved hepatic segments were resected from patients with a suspected diagnosis of cystadenoma. Hepatic resection included segmental resection ($n = 2$ patients), left lateral sectionectomy ($n = 1$), right hemihepatectomy ($n = 2$), and right trisectionectomy ($n = 1$). Both patients with cystadenocarcinoma had been followed by ultrasound imaging elsewhere for several years with the assumption that they had simple, solitary liver cysts. Only one patient with a 14-cm cyst was operable at presentation (right trisectionectomy); the other was found to have widespread hepatic metastases on CT scanning that precluded resection.

One patient with a diagnosis of cystadenoma died during surgery from cardiac arrhythmia and failure. A subphrenic collection complicated liver resection for a cystadenocarcinoma and was successfully treated with percutaneous drainage and antibiotics. There has been no evidence of disease recurrence 61 to 68 months following successful resection (median follow-up 60 months).

Left hemihepatectomy was required in a 23-year-old man with a large, rapidly expanding, painful cyst that had features of recent hemorrhage into the "cyst" and peritoneal cavity. Histology revealed a diagnosis of malignant cystic hepatoblastoma (embryonic sarcoma). The patient received postoperative adjuvant chemotherapy and remains disease-free at 14 months' follow-up.

Discussion

Simple cysts, the commonest cystic lesions of the liver, occurred in 37% of patients in the current report. The diagnosis of simple "uncomplicated" liver cyst(s) can be readily established by cross-sectional imaging. These patients often lack significant symptoms and require no active treatment. No intervention was required in half of our patients with simple cysts. None of the 11 patients with asymptomatic or mildly symptomatic simple cysts became markedly symptomatic or required intervention during a median follow-up of 28 months. These patients may therefore be discharged from follow-up once the simple nature of the cyst(s) has been established radiologically.

More recently, minimally invasive therapeutic approaches have played an increasing role in the management of cystic diseases of the liver. Percutaneous aspiration of symptomatic simple cysts resulted in relief of symptoms in most of our patients but was followed by recurrence of the cyst(s) and symptoms in approximately half of the patients. High recurrence rates following radiologic drainage of simple cysts were also reported by others [8, 9]. The installation of a sclerosant agent such as tetracycline or alcohol may reduce recurrence [10–12]; long-term data remain sparse, however, which has not been helpful in our experience. Nonetheless, percutaneous aspiration may provide temporary symptomatic relief in patients with polycystic disease, may alleviate biliary obstruction, and may help determine whether the cysts are symptomatic [13].

Previous authors suggested that laparoscopic deroofing was particularly useful for management of patients with a large symptomatic solitary cyst [9]. We have favored laparoscopic resection of such lesions whenever possible (see below) to reduce the likelihood of cyst recurrence. Laparoscopic deroofing may be of value in patients with symptomatic multiple simple liver cysts to treat a dominant large cyst. We have employed laparoscopic deroofing in two such patients with good initial results. Symptomatic enlargement of the residual smaller cysts in both patients was managed successfully by further laparoscopic deroofing. This was facilitated by the paucity of adhesions following the initial laparoscopic procedure, a beneficial feature of laparoscopic surgery [14]. Laparoscopic fenestration appears to confer little or no benefit in patients with symptomatic APLD except perhaps for those with large cysts located on the anterior surface of the liver [15, 16]. Our experience with this approach in one patient was rather unfavorable, with rapid deterioration in hepatic function that necessitated rescue liver transplantation. Uncertainty regarding the nature of the cyst, especially when neoplastic pathology is suspected, should be considered an absolute contraindication to laparoscopic deroofing.

Laparoscopic resection of liver cysts and laparoscopic partial hemihepatectomy are alternative minimally invasive approaches to their management [16, 17]. We carried out an uncomplicated laparoscopic partial hemihepatectomy in three patients with solitary cysts that replaced most of the resected hepatic segments. Cysts located in the left lateral section of the liver (segments II and III) are best suited technically for this approach. Laparoscopic resection remained feasible when segment IV was also affected, but this involvement added difficulty to the procedure owing to the greater thickness of hepatic tissue to be divided and the larger number of vascular structures and biliary radicles to be ligated. Technologic advances, such as the ultrasonic dissector

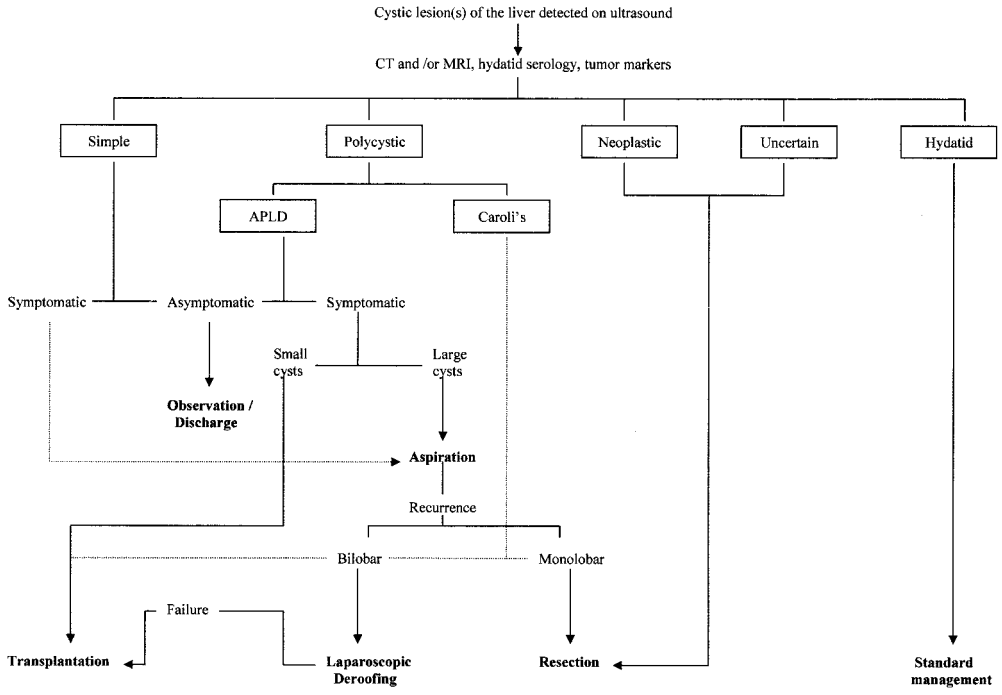

Fig. 5. Management strategy for patients with nonparasitic cystic lesions of the liver. APLD: adult polycystic liver disease.

(Harmonic Scalpel), may facilitate laparoscopic resection and eliminate the need for ligation [18, 19], though we have had no experience with their use. Hepatic resection ensures complete excision of the cyst wall, which may reduce the likelihood of cyst recurrence. However, other small (< 1.5 cm in diameter), asymptomatic cysts developed in the remaining liver in two patients during the initial 2 years of follow-up, which reflects the often-diffuse nature of this pathology. Careful long-term evaluation of the efficacy of laparoscopic techniques for management of cystic liver disease is required.

Despite the diminishing role of open surgery in the management of simple hepatic cysts, open hepatic resection remains the mainstay of surgical therapy for managing neoplastic cysts of the liver [20]. Patients with symptomatic Caroli's disease who have a monolobar disease may also be managed successfully by hepatic resection [21–23]. We obtained good results in two of three patients with unilobar Caroli's disease that we treated by liver resection. Attention should be given to the clearance of intrahepatic calculi, which can be extensive in these patients [24]. Progression of disease to involve the remaining liver should be managed by liver transplantation.

Although a more extensive resection with hepaticojejunostomy is occasionally possible in patients who present with bilobar Caroli's disease [25], our limited experience with this approach was not favorable, and such patients may be managed by liver transplantation in the first instance. Disease progression in the remaining liver and the considerable risk of malignant transformation [26, 27] are major considerations. Cholangiocarcinoma has also been reported in a patient with monolobar Caroli's disease [28]. Cholangiocarcinoma, the commonest malignancy in patients with Caroli's disease, was previously reported in 7% to 14% of patients [26, 29]. In the current series, cholangiocarcinoma occurred in three of eight patients treated at our unit (37.5%). It was detected in two of five patients who presented with bilobar involvement and in one of three patients with unilobar involvement (treated by resection) and subsequent cystic degeneration in the remaining liver. Hepatocellular carcinoma, undifferentiated carcinoma, and squamous cell carcinoma have also been reported [30–32].

Although the alarmingly high incidence of malignancy in these patients calls for constant surveillance, there are currently no reliable methods for early detection of cholangiocarcinoma in these patients. In two of the three patients with malignant transformation, the detection of cholangiocarcinoma in the explanted livers was unsuspected. However, fine-needle aspiration cytology and ultrasound-guided biopsy of papillary lesions within biliary cysts have been used by others [29, 31] for this purpose with some success, and the biliary tumor marker CA19-9 may be of some benefit. In the absence of reliable indicators of biliary dysplasia and early cholangiocarcinoma, and taking into consideration the dismal prognosis of cholangiocarcinoma once established [33], early decision to transplant patients with bilobar disease is emphasized.

Patients with liver cysts of uncertain etiology should undergo liver resection. The preoperative diagnosis may be enhanced by the detection of elevated serum concentrations of the tumor marker CA 19-9, which often suggests cystadenoma or cystadenocarcinoma [34, 35]. Percutaneous aspiration of cyst contents for cytologic analysis, however, should be avoided, as it may lead to intraperitoneal seeding or needle track implantation from an undiagnosed cystic neoplasm of the liver [36, 37]. Although most cystic lesions that mimic neoplasms of the liver prove after resection to be simple cysts complicated by hemorrhage or infection, about one-fourth of them are neoplastic [38]. In our series, one of four patients with suspicious liver cysts was shown to have an embryonic sarcoma (hepatoblastoma) on histology; the remaining three had complicated simple cysts.

Some patients with APLD may also benefit from partial hemihepatectomy [39–41], as was the case for one of our patients

treated by right hemihepatectomy. This approach should be considered carefully, as postresection liver failure and death may ensue [40, 42]. This risk may not be reduced by laparoscopic surgery. Indeed, one patient in our series had to be given a transplant following laparoscopic fenestration for deteriorating liver function. Moreover, recurrence of symptoms following resection or fenestration (or both) is not uncommon [41].

Total hemihepatectomy and orthotopic liver transplantation may be considered in APLD patients with intractable abdominal pain and distension. Hepatocellular failure is rarely observed in these patients. Combined liver and kidney transplantation may become the treatment of choice in patients with concomitant renal involvement and worsening renal function. Careful assessment of renal function in these patients is therefore necessary. Though the experience with transplantation for APLD is limited, it appears to achieve excellent palliation. Good results were obtained in 14 of 17 patients reported in three series [43–45]; the remaining 3 patients died perioperatively. Liver transplantation resulted in resolution of symptoms in four of the five patients receiving transplants in our series; the fifth patient died. The decision to give transplants to patients with APLD calls for fine judgment on the part of the physician and surgeon involved and should take into account the potential benefits of the procedure and the risks associated with surgery and immunosuppression, as well as the current shortage in organ availability.

Although no treatment is required in one-third of patients with benign cystic liver disease, radiologic and surgical interventions are often indicated. Although recurrence of cyst and symptoms is common after radiologic treatment of patients with simple cysts or APLD, this approach may be reserved for those who refuse, or who are unfit for, surgery and to confirm the potential for symptomatic response prior to embarking on surgery. Laparoscopic cyst fenestration may be best reserved for patients with solitary simple cysts, though hepatic resection may be preferable, as it minimizes the chances of recurrence. Patients with unilobar Caroli's disease can be treated with resection. Progression of disease and bilateral involvement call for early consideration of liver transplantation, as the risk of malignant transformation is considerable. In contrast, liver transplantation is a reserved option in patients with extensive APLD that requires careful consideration of the benefits and risks, as these patients often have adequate hepatic reserve. Patients with solitary liver cysts of uncertain etiology should be considered for liver resection, as one-fourth of them might be malignant. An algorithm for the management of cystic lesion(s) of the liver is proposed (Fig. 5).

Résumé. Dans ce travail on définit les indications et on évalue les résultats de différentes options thérapeutiques des patients porteurs d'une lésion kystique du foie. Entre 1992 et 1999, nous avons traité 60 patients consécutifs porteurs d'une maladie kystique du foie. Le diagnostic a été un kyste simple (solitarie 12 fois, multiple 10 fois), la maladie polykystique de l'adulte (MPKA; $n = 17$), la maladie de Caroli ($n = 8$), un kyste hydatique ($n = 4$) et les kystes néoplasiques ($n = 9$). La moitié des patients porteurs de kyste simple avaient des symptômes inexistants ou modérés et ne nécessitaient aucun traitement. On a effectué un drainage par voie percutanée chez huit patients (kyste simple; $n = 4$, MPKA; $n = 4$), suivi de récidive symptomatique chez trois. On a réalisé une résection du dôme saillant sous laparoscopie chez trois patients (kystes simples multiples; $n = 2$, MPKA; $n = 1$), suivi de grossissement symptomatique des kystes restants nécessitant une intervention ultérieure (résection du dôme saillant; $n = 2$ et transplantation; $n = 1$). Une hépatectomie a été réalisée sous coelioscopie chez trois patients avec des kystes solitaires simples. Des 18 patients ayant eu une résection hépatique par laparotomie (kyste néoplasique; $n = 8$, maladie de Caroli; $n = 4$; kyste simple; $n = 3$, kyste hydatique; $n = 2$ et MPKA; $n = 1$), deux patients, porteurs de maladie de Caroli, ont nécessité une transplantation hépatique pour maladie progressive. Neuf patients (maladie de Caroli; $n = 5$, MPKA; $n = 4$) ont eu une transplantation du foie, dont trois avec une transplantation concomitante du rein. Sept patients ont développé des complications et trois en sont décédés (5%). Trois patients, porteurs de maladie de Caroli bilatérale, ont développé un cholangiocarcinome et en sont décédés. Le traitement radiologique interventionnel a un rôle limité dans le traitement des patients atteints de kyste simple ou de MPKA. On peut réaliser la résection du dôme saillant sous coelioscopie des kystes simples, éventuellement de façon itérative, mais la résection minimise le risque de récidive. La maladie de Caroli unilobaire peut être traitée par résection mais la forme bilatérale nécessite une transplantation précoce pour éviter le risque de transformation maligne. La transplantation a des indications limitées chez le patient porteur de MPKA.

Resumen. Este artículo define las indicaciones y evalúa los resultados de las diferentes opciones de manejo en pacientes con enfermedades quísticas del hígado, con base en 60 pacientes consecutivos tratados en el periodo entre 1992 y 1999. Los diagnósticos incluyeron quiste simple (solitario 12, múltiple 10), enfermedad poliquística hepática del adulto (EPHA, 17), enfermedad de Caroli (8), quiste hidatídico (4), y quistes neoplásicos (9). La mitad de los pacientes con quistes simples exhibieron síntomas leves o fueron asintomáticos y no requirieron tratamiento. El drenaje percutáneo fue practicado en 8 pacientes (quiste simple 4, EPHA 4) y se presentó recurrencia sintomática en 3. El "destechamiento" (de roofing) del quiste se practicó en 3 pacientes (quistes simples múltiples 2, EPHA 1) y fue seguido de aumento sintomático del tamaño de los quistes remanentes que requirió intervención adicional ("destechamiento" laparoscópico 2, trasplante 1). La hepatectomía laparoscópica resultó exitosa en 3 pacientes con quistes simples solitarios. De 18 pacientes sometidos a resección hepática abierta (neoplásica 8, Caroli 4, quistes simples 3, quiste hidatídico 2, APHA 1), dos pacientes con enfermedad de Caroli requirieron trasplante de hígado por progresión de la enfermedad; 9 pacientes (Caroli 5, APHA 4) recibieron trasplante hepático y 3 tuvieron trasplante renal concomitante; 7 pacientes desarrollaron complicaciones y 3 murieron (5%). Se desarrolló colangiocarcinoma en tres pacientes con enfermedad de Caroli bilateral y todos murieron. El tratamiento radiológico tiene un papel limitado en el manejo de pacientes con quistes simples o con APHA. El "destechamiento" de los quistes simples debe ser repetido, en tanto que la resección minimiza la recurrencia. La enfermedad de Caroli unilobar puede ser resecada, en tanto que la enfermedad bilateral requiere trasplante hepático temprano debido al alto riesgo de malignidad. El trasplante se reserva como modalidad terapéutica para pacientes con APHA extensa.

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