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Adult Polycystic Liver Disease

Is Fenestration the Most Adequate Operation for Long-Term Management?

Jean-François Gigot, M.D.,* Pascale Jadoul, M.D.,* Florencia Que, M.D.,†
Bernard E. Van Beers, M.D.‡, José Etienne, M.D.,* Yves Horsmans, M.D.,§
Alexandra Collard, M.D.,* André Geubel, M.D.,§ Jacques Pringot, M.D.,‡
and Paul-Jacques Kestens, M.D.*

From the Departments of Digestive Surgery, Medical Imaging,‡ and Gastro-Enterology,§ St-Luc University Hospital, Louvain Medical School, Brussels, Belgium; and the Department of Gastroenterologic and General Surgery,† Mayo Clinic and Mayo Foundation, Rochester, Minnesota*

Objective
The aim of this study was to evaluate the immediate and long-term results in a retrospective series of patients with highly symptomatic adult polycystic liver disease (APLD) treated by extensive fenestration techniques. A classification of APLD was developed as a stratification scheme to help surgeons conceptualize which operation to offer to patients with APLD.

Summary Background Data
Treatment options for APLD remain controversial, with partisans of fenestration techniques or combined liver resection-fenestration.

Methods
Clinical symptoms, performance status, liver volume measurement by computed tomography (CT), and morbidity were recorded before surgery and after surgery. Adult polycystic liver disease was classified according to the number, size, and location of liver cysts and the amount of remaining liver parenchyma. Follow-up was obtained by clinical and CT examinations in all patients.

Results
Ten patients with highly symptomatic APLD were operated on using an extensive fenestration technique (by laparotomy in 8 patients and by laparoscopy in 2 patients, 1 of whom conversion to laparotomy was required). The mean preoperative liver volume was 7761 cm^3. There was no mortality. Postoperative morbidity occurred in 50%, mainly from biliary complications, requiring reintervention in two cases. Massive intraoperative hemorrhage occurred in one patient. During a mean follow-up time of 71 months (range, 17 to 239 months), all patients were improved clinically according to their estimated performance status. The mean postoperative liver volume was 4596 cm^3, which represents a mean liver volume reduction rate of 43%. However, in type III APLD, despite absence of clinical symptoms, a significant increase in liver volume was observed in 40% of the patients.

Conclusions

Extensive fenestration is effective in relieving symptoms in patients with APLD. Hemorrhage and biliary complications are possible consequences of such an aggressive attempt to reduce liver volume. The procedure can be performed laparoscopically in type I APLD. A longer follow-up period is mandatory in type II APLD, to confirm the usefulness of the fenestration procedure. In type III APLD, significant disease progression was observed in 40% of the patients during long-term follow-up. Fenestration may not be the most appropriate operation for long-term management of all types of APLD.

Adult polycystic liver disease (APLD) is a rare affliction with a prevalence in autopsy series between 0.08% and 0.53%.[1-4] Frequently associated with autosomal dominant polycystic kidney disease,[1-8] APLD generally is asymptomatic and does not then require surgical treatment. However, a number of patients with highly symptomatic cystic hepatomegaly or with a complicated presentation will benefit from surgical decompression of the hepatomegaly.[6] Currently, the most appropriate therapeutic approach for APLD remains controversial. Transient improvement with nonsurgical treatment such as aspiration or ablation with alcohol or sclerosing agents has been reported.[9-11] The surgical approach has partisans of the fenestration procedure[12-19] with or without liver resection,[20-29] or liver transplantation.[9,30,31]

The purpose of this study was to evaluate the immediate and long-term outcomes of patients with highly symptomatic APLD treated surgically by fenestration techniques in a 19-year period at a single institution. We attempted to develop a stratification scheme based on classification of the type of APLD to help surgeons conceptualize which operation to offer to these patients.

PATIENTS AND METHODS

Between November 1975 and May 1994, 14 patients suffering from symptomatic APLD were referred to the surgical department of St-Luc University hospital in Brussels. Ten patients with highly symptomatic APLD were selected for surgical treatment. Four patients with minimally symptomatic APLD were treated conservatively and were excluded from this series.

There were nine women and one man, with a mean age of 47 years (median, 48; range, 30 to 61 years). Five patients had a familial history of APLD and five patients of associated autosomal dominant polycystic kidney disease. The average time between diagnosis of APLD and surgery was 8 years (median, 9; range, 1 to 18 years). The mean duration of symptoms was 58 months (median, 36; range, 12 to 180 months). Chronic abdominal pain was present in all patients, as reported solely in one and associated with incapacitating abdominal fullness in eight, dyspepsia in five, leg edema in three, supine dyspnea in two, and elevated temperature related to a superinfected cyst in one. The estimated preoperative American Cancer Society Eastern Cooperative Oncology Group (ECOG) performance status score[32] was grade 3 in one patient, 2 in four patients, 1 in four patients, and 0 in one patient (the one with a superinfected cyst). At abdominal examination, hepatomegaly and abdominal distention were present in all patients. Five patients had previous treatment of APLD, including alcohol injection in four patients (mean number, 2; range, 2 to 4 injections) and incomplete cyst fenestration by laparotomy in one patient. Preoperative liver function test results were normal in four patients, but slight elevation of alkaline phosphatase (mean, 1.4; median, 1.1; range, 1.0 to 2.2 × normal values) and γ-glutamyl transferase (mean, 3.4; median, 3; range, 1.6 to 4.8 × normal values) was present in the remaining patients. Serum creatinine and coagulation test results were normal in all patients. Imaging studies included abdominal ultrasound and computed tomography (CT) in all but one patient who was operated on in 1975 when preoperative CT was not available. We used a personal classification of APLD according to the number and size of liver cysts and the amount of remaining liver parenchyma. *Type I* included patients with a limited number (<10) of large cysts (>10 cm) (Fig. 1A). *Type II* was represented by patients with diffuse involvement of liver parenchyma by multiple medium-sized cysts with remaining large areas of noncystic liver parenchyma on preoperative CT (Fig. 1B). *Type III* was a severe form of APLD with massive, diffuse involvement of liver parenchyma by small- and medium-sized liver cysts and only a few areas of normal liver parenchyma between cysts (Fig. 1C). Liver volume was calculated by using the public domain National Institutes of Health Image Software (Bethesda, MD)[21,33] from preoperative and serial postoperative abdominal CT. One patient presented with an asymptomatic small intracranial aneurysm detected by routine high-resolution CT scanning.[34,35]

The operative approach used in this series was open laparotomy in eight patients (through a bilateral subcostal

Address reprint requests to Jean-François Gigot, M.D., Department of Digestive Surgery, St-Luc University Hospital, Hippocrate Avenue 10, B - 1200, Brussels, Belgium.

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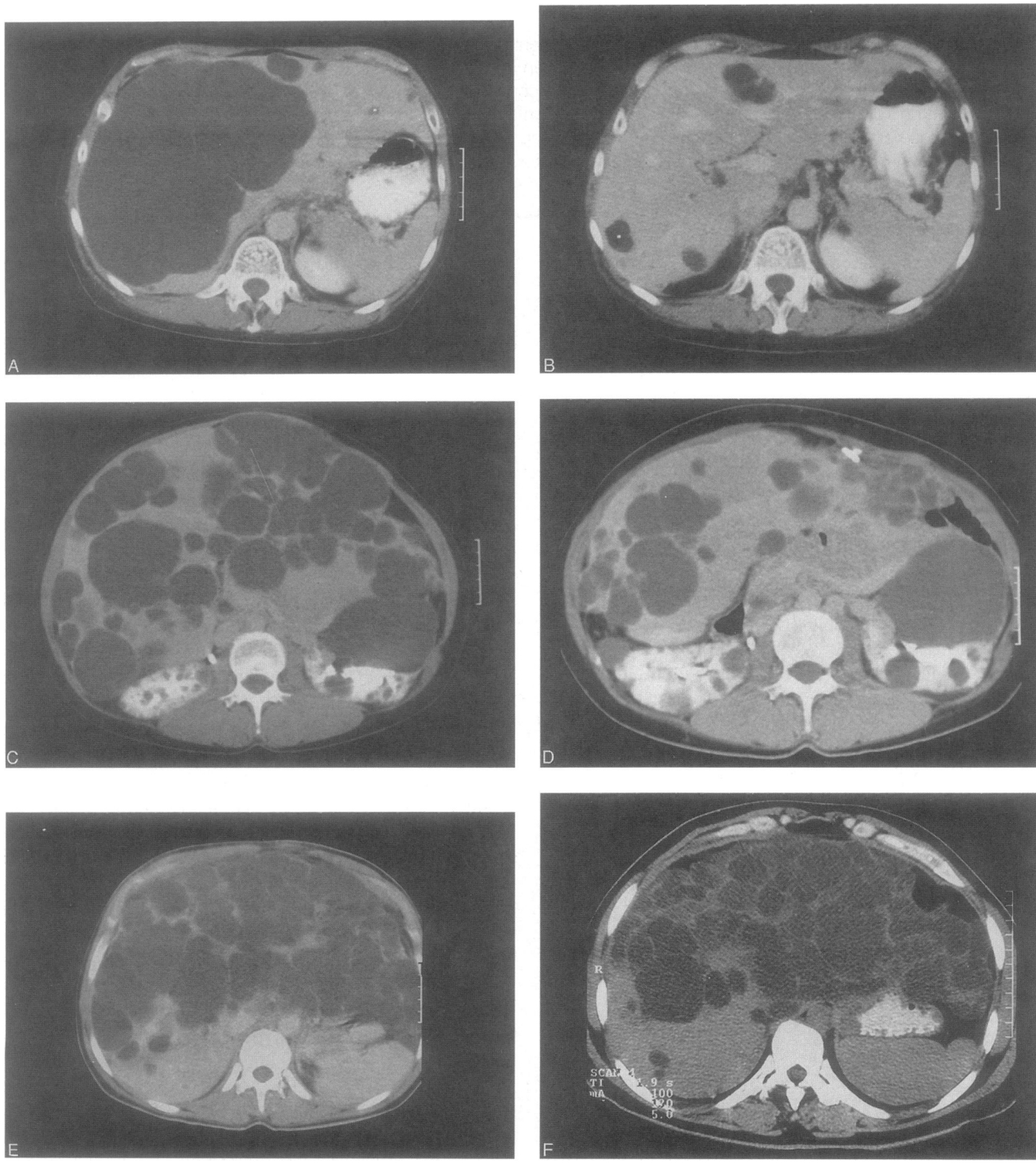

Figure 1. Suggested classification of patients with adult polycystic liver disease (APLD) according to number, size, and location of cysts within the liver, and the amount of residual normal liver parenchyma on preoperative computed tomography (CT). (A) Type I APLD. Limited number (<10) of large cysts (>10 cm). Preoperative CT and postoperative CT (B). (B) Type II APLD. Diffuse involvement of liver parenchyma by multiple medium-sized cysts with remaining large areas of noncystic liver parenchyma. Preoperative CT (C), postoperative CT (D). (C) Type III APLD. The most severe form of APLD with massive, diffuse involvement of liver parenchyma by small- and medium-sized liver cysts, with only a few area of remaining normal liver parenchyma between cysts. Preoperative CT (E) and postoperative CT (F).

incision) and a laparoscopic approach (using a 4-trocar technique) in two patients. The fenestration technique described by Lin et al.[12] was applied in all patients, with deroofing of as many cysts as possible, starting from the superficial and then, stepwise, opening the deep-sited cysts with the help of intraoperative ultrasound. Great care was taken to avoid vascular and biliary tract injury within the cystic septa. The average volume of fluid aspirated during fenestration was recorded. Cholecystectomy was performed as part of the initial surgical procedure in three patients, allowing routine control intraoperative cholangiography and methylene blue dye biliary injection, after completion of the procedure, to detect any inadvertent intracystic biliary leak. When leakage occurred (two patients), choledochotomy with routine T-tube biliary drainage was used in addition to biliary leak suture. Peritoneal drainage was used in eight patients.

Postoperative clinical evaluation was performed by follow-up visit in all patients. The mean follow-up time was 71 months (median, 55 months; range, 17 to 239 months) in the entire series. Criteria used for postoperative clinical evaluation included postoperative morbidity, improvement or reappearance of symptoms, and estimated ECOG performance status. Postoperative morphologic evaluation included repeated abdominal CT with liver volume measurement. The mean number of CT examinations per patient was 5 (median, 4; range, 1 to 13 examinations). Statistical analysis included appropriate use of chi square test or Student's t test when indicated.

RESULTS

Preoperative Features and Surgical Approach

One patient had type I APLD, four patients had type II, and five patients had type III. The mean age of the patients, the mode of presentation, the delay between diagnosis of APLD and surgery, and the duration of symptoms were not significantly different among the three types of APLD. Preoperative estimated ECOG performance status was grade 1 in the patient with type I APLD; grade 3 in one patient, grade 1 in two patients, and grade 0 in one patient with type II; and grade 2 in four patients and grade 1 in one patient with type III. Preoperative liver volume was calculated in only nine patients because the remaining patient had been operated on in 1975, at which time preoperative CT was not available. The mean preoperative liver volume was 7761 cm^3 (median, 7576; range, 4200 to 10,956 cm^3). The preoperative liver volume was greater in type III compared with type II APLD, but the difference was not statistically significant. The patient with type I APLD underwent successful laparoscopic fenestration, allowing aspiration of 2.5 L of fluid during an operative time of 120 minutes. The four patients with type II APLD underwent open fenestration with a mean operative time of 220 minutes (median, 215; range, 180 to 270 minutes). The average volume of fluid aspirated at laparotomy recorded from the operative records in three of these patients was 3.3 L (median, 2.5; range, 2.5 to 5.0 L). The mean volume of blood or other liquid replacement in this group was 320 mL (median, 140; range, 0 to 1000 mL) and 2210 mL (median, 1920; range, 1000 to 4000 mL), respectively. Of the five patients with type III APLD, one was found to be poorly suited for laparoscopic fenestration and required conversion to laparotomy because of incomplete fenestration. The average operative time in this group was 364 minutes (median, 360; range, 250 to 480 minutes), and the average volume of fluid aspirated at laparotomy, recorded from the operative records in all patients, was 8.6 L (median, 7; range, 5 to 15 L). Except in one patient of this group who had massive intraoperative hemorrhage requiring 34 units of blood, the mean volume of blood or other liquid replacement in the remaining four patients was 998 mL (median, 1006; range, 0 to 1977 mL) and 4400 mL (median, 3400 mL; range, 3200 to 6000 mL), respectively. The mean operative time ($p < 0.008$) and the mean volume of fluid aspirated during fenestration ($p < 0.04$) were significantly greater in patients with type III APLD. The overall volume of blood and other liquid replacement during surgery was not significantly different.

Intraoperative and Postoperative Morbidity

There was no intraoperative or postoperative mortality in this series. The patient with successful laparoscopic fenestration had an uncomplicated postoperative course and was discharged 7 days after surgery. During open surgery, two major complications were encountered. One was a massive hemorrhage in a patient with type III APLD, associated with diffuse intravascular coagulopathy. The hemorrhage was related to accidental injury of the right hepatic vein during fenestration of deep posterior liver cysts after tedious mobilization of the right posterior liver segment. In another patient, with type II APLD, instrumental injury to the right posterolateral bile duct occurred during interseptal fenestration of right posterior deep-sited liver cysts. This biliary tear was recognized adequately and was treated by primary suture under T-tube protection. Long-term outcome was satisfactory in this patient.

The postoperative course was uncomplicated in two patients (50%) with type II APLD and in two patients (40%) with type III APLD (not significant). Biliary complications occurred in three patients (Fig. 2) (one patient with type II APLD and two patients with type III APLD)

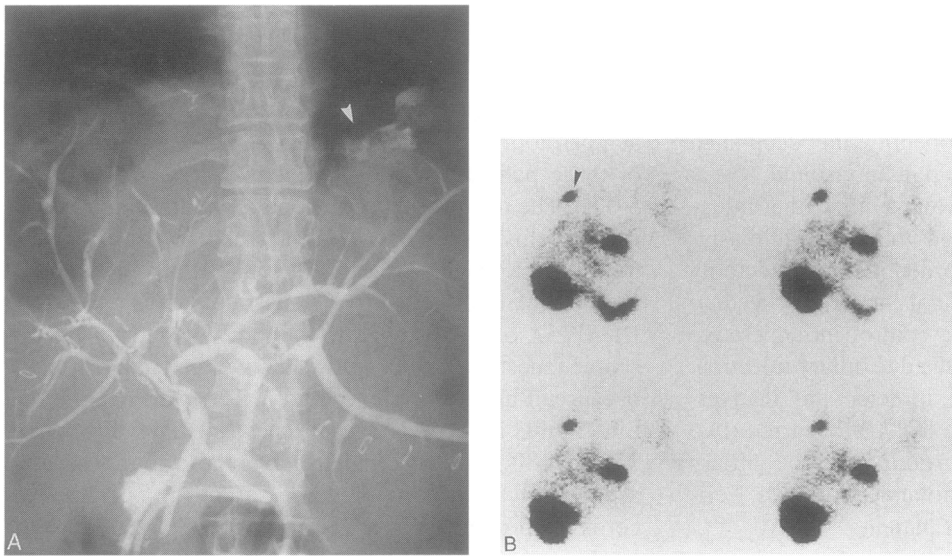

Figure 2. Biliary complications during open fenestration procedure in patients with adult polycystic liver disease. (A) Intraoperative cholangiogram shows contrast extravasation (arrow) within a left liver cyst after open fenestration. (B) Postoperative technetium 99m-iminodiacetic acid biliary scintigraphy shows localized extravasation in one liver cyst after open fenestration in another patient. Spontaneous resolution occurred in this case.

without operative biliary exploration. Two patients had to undergo reoperation for bile ascites in one case (without peritoneal drainage) and for persistent biliary leak in the other (with peritoneal drainage). Reoperation included intracystic biliary leak suture, choledochotomy, and T-tube placement in both cases. In the patient with massive intraoperative hemorrhage, small obstructive pigmentary stones were responsible for postoperative obstructive jaundice and were removed during reoperation and common bile duct exploration. The same patient had a persistent ascitic leak, which resolved within 2 weeks. The third patient with biliary complication had a postoperative asymptomatic biliocystic fistula detected on postoperative T-tube cholangiogram and technetium 99m-iminodiacetic acid biliary scintigraphy. This patient had spontaneous resolution as noted on subsequent radiologic examination, and the T-tube was removed 2 months after the initial surgery. A final patient with type III APLD (who was limited to a standing position before surgery) had an inferior vena caval compression syndrome with bilateral leg edema during the early postoperative period. This complication resolved without recurrence with medical treatment and ambulatory rehabilitation. The mean postoperative stay of all patients in this series was 18 days (median, 14.5; range, 7 to 43 days). For patients with an uncomplicated postoperative course, the mean postoperative stay was 9.2 days (median, 9; range, 7 to 12 days) compared with 27 days for patients with postoperative complications (median, 25; range, 17 to 43 days) ($p < 0.004$).

Long-Term Clinical Results

The patient with type I APLD who was treated laparoscopically remained free of symptoms, with normal liver function test results and without cyst recurrence 56 months after surgery. During a mean clinical follow-up of 83 months (median, 38 months; range, 17 to 239 months), all four patients with type II APLD remained free of symptoms, with normal liver function test results and with postoperative estimated ECOG performance status grades of 0 in all patients. During a mean clinical follow-up of 65 months (median, 60; range, 27 to 94 months), one patient with type III APLD had recurrent epigastric pain related to an increase in size of a liver cyst in segment IV of the liver. This patient was treated successfully 12 months after initial fenestration by intracystic alcohol cyst ablation. One patient had normal liver function test results, slight elevations of γ-glutamyl transferase on two occasions (less than 3 times the normal value), and two patients had significantly increased values of alkaline phosphatase (3 and 6 $\times$ normal values) and of γ-glutamyl transferase (8 and 19 $\times$ normal values). Bilirubin level was normal in all patients. The two patients with abnormal liver function tests remained free of symptoms. The postoperative estimated ECOG performance status in patients with type III APLD was graded 1 in two patients and graded 0 in three patients.

Long-Term Morphologic Results

Repeated liver CT examinations were performed on nine patients during a mean radiologic follow-up time of 49 months (median, 51; range, 10 to 91 months). One patient with type II APLD had only preoperative liver ultrasound, and thus comparison of preoperative and postoperative liver volumes was not available. Subjective examination results of postoperative liver CT were disappointing in patients with types II and III APLD, despite

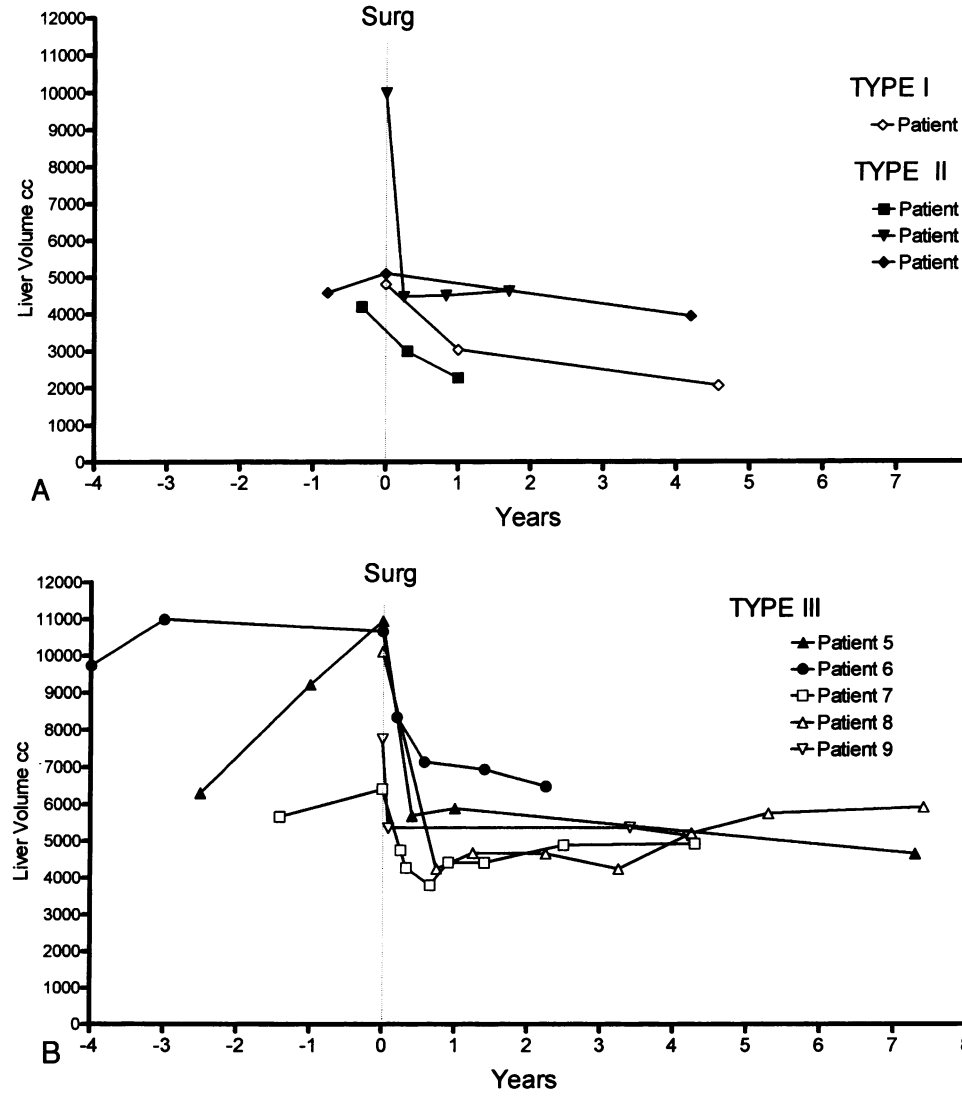

Figure 3. Evaluation of liver volume reduction between preoperative and serial postoperative liver computed tomographic examination during follow-up of patients operated on by fenestration for adult polycystic liver disease (APLD). (A) Profile of preoperative and sequential postoperative evolution in liver volume during follow-up in patients with type I and II APLD. (B) Profile of preoperative and sequential postoperative evolution in liver volume during follow-up in patients with type III APLD.

an overall impression of a decrease in the number and size of cysts within the liver. Objective CT evaluation results at 6 months showed an average postoperative liver volume of 4596 cm^3 (median, 4364; range, 3001 to 6049 cm^3), which represents a mean reduction in liver volume of 41% (median, 39; range, 29% to 58%) compared with preoperative liver volume. During radiologic follow-up (mean duration, 49 months), the average postoperative liver volume was 4450 cm^3 (median, 4658; range, 2065 to 6480 cm^3), which represents an average liver volume reduction of 43% (median, 42%; range, 23% to 57%) compared with preoperative liver volume. The comparative duration of follow-up, the mean preoperative and postoperative liver volumes, and the reduction in liver volume in the various types of APLD are detailed in Table 1. The mean follow-up time for radiologic (CT) follow-up was shorter in patients with type II APLD than in those with type III APLD. There was no significant difference regarding liver volume reduction after fenestration between patients with type II and type III APLD. However, all patients with type I or II APLD experienced stable reduction in liver volume during the follow-up time (Fig. 3A) compared with patients with type III APLD (Fig. 3B). Two patients (40%) with type III APLD experienced a significant increase in liver volume, of 30% and 39% compared with the 6 months postoperative liver volume at a follow-up time of 52 and 89 months, respectively (patients 7 and 8). In these two patients, who remained free of symptoms, disease progression seemed mainly related to an increase in size of pre-existing untreated deep liver cysts.

DISCUSSION

In patients with APLD, there is some controversy over the optimal treatment, which includes nonsurgical ap-

Table 1. REDUCTION IN LIVER VOLUME DURING RADIOLOGIC (COMPUTED TOMOGRAPHY) FOLLOW-UP IN PATIENTS WITH ADULT POLYCYSTIC LIVER DISEASE (APLD) ACCORDING TO TYPE OF APLD

Type of APLD	No. of Patients	Mean Preoperative Liver Volume (cm^3)	Liver Volume at 6-Month Follow-up		Mean Duration of Radiologic Follow-up (mo)	Liver Volume at Final Long-Term Follow-Up	
			Mean Volume (cm^3)	Mean Reduction Rate (%)		Mean Volume (cm^3)	Mean Reduction Rate (%)
I	1	4816	3046	37	55	2065	57
II	3	6435	3738	42	28	3617	44
III	5	9146	5249	43	61	5427	41

proaches (*e.g.,* intracystic alcohol injection) or surgical approaches, by laparotomy or more recently by laparoscopy.[36–38] Regarding surgical treatment, controversy persists between "conservative" fenestration[12–19] and combined hepatic resection-fenestration,[20–29] which involves significant mortality and morbidity.[21] Even extensive fenestration must be considered as a formidable operation, as proved in our series and reported by Turnage et al.[26] Extensive hepatic surgery in these patients should therefore be restricted to those with highly symptomatic polycystic hepatomegaly that seriously impairs their quality of life or those with complicated disease such as biliary compression,[17,19,26,39–43] portal hypertension,[8,17,44–47] hepatic venous outflow obstruction,[48–50] inferior vena caval compression,[26,51] liver cyst superinfection,[26,36,52–57] or cancer.[36,58–60] Postoperative morbidity mainly is related to hemorrhage and biliary complications. Hemorrhage is the more common cause of intraoperative or postoperative death in surgical cases.[17,26] The most dangerous phase of the fenestration procedure is unroofing of right deep-sited liver cysts. Adequate complete mobilization of the right lobe in massive polycystic hepatomegaly can be difficult and tedious, and the position and direction of the right hepatic vein usually are distorted. This was the mechanism for the massive intraoperative hemorrhage in one of our patients. Urgent attempt at blind hemostatic control of the hepatic vein also can lead to hepatic outflow obstruction.[61] The same risk of injury is encountered with biliary ducts, which also are distorted in polycystic liver anatomy.[62] They can be injured easily within cystic septa during the fenestration procedure. Biliary complications represent the major problem for us during fenestration procedures in APLD. For this reason, we advocate routine intraoperative cholangiography and methylene blue dye biliary injection under pressure to detect any biliary leaks within the fenestrated interseptal cystic wall. Primary suture and T-tube drainage allowed successful conservative treatment of such a complication in one of our last patients. Biliary complications occurred in our series in patients with either type II or type III APLD and were related in both types to interseptal fenestration for deep-sited liver cysts. Our approach to this problem was the use of preoperative liver CT and intraoperative ultrasound to locate the deep-sited cysts and to choose the best approach for interseptal cystic fenestration.

The final purpose of surgical treatment in APLD is to reduce significantly the size of severe polycystic hepatomegaly and to provide long-term relief of symptoms. Impressive postoperative reduction in liver volume (62%) has been reported in patients treated by liver resection-fenestration.[21] We believe ours is the first surgical series in which the same objective criteria[21,33] were used to evaluate liver volume reduction in patients with APLD treated by fenestration without liver resection. The inferior reduction in liver volume after fenestration in patients with type II compared with type III APLD could be considered surprising. However, the presence of a large remaining area of normal liver parenchyma between liver cysts, especially between superficial and deep-sited cysts, could explain the difficulty of unroofing deep-sited liver cysts and, when aggressive attempts are made, the increased risk of biliary complications. The most important question is the risk of disease progression during extended follow-up time to 20 to 30 years. The potential deterioration of results with fenestration procedures in these type II patients is unknown, whereas during a 5-year follow-up, significant disease progression already was observed in 40% of the patients with type III APLD, mostly related to a progressive increase in size of deep residual untreated liver cysts rather than new cyst development.

Finally, we propose a stratification scheme for the most appropriate surgical approach in patients with APLD, based on liver cystic and parenchymal liver anatomy as determined by preoperative CT. Type I APLD, which represents only 10% of our series, already has been stated by Morino et al.[37] to be an excellent indication for laparoscopic fenestration. Type III APLD represents the most severe form of the disease, and these patients already have been identified in the Bismuth experience[18] as having poor results. Our experience suggests that even aggressive

fenestration is associated with disease progression in this particular subgroup of patients. Despite extensive fenestration, a huge skeletal architecture of fenestrated cystic wall will remain and impair liver volume reduction. These patients are thus the best candidates for combined liver resection-fenestration. The rate of reduction in liver volume achieved in patients with type II APLD after extensive fenestration in this series was excellent and stable, although we cannot yet exclude long-term disease progression. We advocate fenestration in this group because liver resection is a more aggressive procedure, which could result in the excision of large amounts of remaining normal liver parenchyma. Conversely, the fenestration technique mostly is efficient for superficial liver cysts and is associated with an increased morbidity when aggressive fenestration of deep-sited cysts is attempted, whereas liver resection enables one to treat superficial and deep-sited cysts, especially when unilobar distribution is present. We presently emphasize the use of the aforementioned classification to stratify the surgical approach in these difficult patients. Liver transplantation should be reserved for patients with liver insufficiency,[21,23] recurrence after initial liver resection,[20,25,28] or associated severe kidney disease,[23,29–31] combining in this case liver and kidney transplantation.

Finally, regarding the difficulty of reoperation, treatment should be performed by surgical teams experienced in the management of these problematic patients.

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