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Polycystic Liver Disease: Experience at a Teaching Hospital

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OBJECTIVES: This study describes the natural history of patients with polycystic liver disease, a rare disorder characterized by multiple hepatic cysts.

METHODS: Cases were identified through review of charts from a hepatology practice, a hepatobiliary surgery practice and a retrospective chart review of inpatient charts from 1990 to 2002. All patients had greater than four simple liver cysts without infectious etiology. Medical records were reviewed for history, physical examination, imaging, and laboratory data. Patients' family practitioners provided follow-up.

RESULTS: Fifty-three cases (62.3% female, 37.7% male) were identified. The mean age at diagnosis was 56.4 yr. Thirty-eight cases (71.7%) had associated polycystic kidney disease. The minority of patients were symptomatic at diagnosis (pain in 19 (36.5%), dyspnea in 5 (9.6%), and restricted mobility in 5 (9.6%) with hepatomegaly in 23 (45.1%). Follow-up information was attainable for 40 patients with a mean follow-up duration of 4.69 yr (range 0.1–15 yr). Within this subgroup, 9 patients (22.5%) had cyst bleeding, 5 (12.5%) had cyst rupture, 5 (12.5%) had cyst infection, 12 (30%) required an intervention. One patient (2.5%) developed portal hypertension, and two (5%) received a liver transplant. One patient (2.5%) died due to complications from liver cysts.

CONCLUSIONS: Most patients in this highly selected cohort were asymptomatic with normal hepatic function. Pain was the most common symptom. The natural history is variable however, with some patients developing complications including portal hypertension. Minimally invasive interventions are appropriate initially, with hepatic resection and liver transplantation reserved for those with severe symptoms or life-threatening complications.

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INTRODUCTION

Polycystic liver disease (PLD) is a rare clinical entity with a prevalence of 0.05–0.13% in autopsy studies (1, 2). It is most commonly associated with autosomal dominant polycystic kidney disease (ADPKD), where the development of hepatic cysts lags behind the onset of renal cysts. The incidence of hepatic cysts in ADPKD ranges from 29% to 48% (2–5) depending on the population examined and the diagnostic method used. The frequency of hepatic cysts in ADPKD increases with the age of the population studied, with a prevalence of 11–27% in patients under age 30 compared to 77–83% over age 60 (3–5).

In addition to the association with ADPKD, an autosomal dominant form of isolated PLD has been described with no relation to mutations in PKD1 or PKD2 (6, 7). Unlike ADPKD, this condition is not associated with renal cysts or intracranial aneurysms (1, 8). The causative gene has been identified on chromosome 19p, which encodes the protein hepatocystin (9, 10). The cysts in both forms of PLD are thought to arise from malformation of the embryonic ductal plate with formation of von Meyenberg complexes (dilated bile ducts in a fibrous stroma, which do not communicate with the biliary tree) (2, 11, 12). The functional biliary epithelium within these complexes secretes fluid causing cyst formation.

In most cases PLD is asymptomatic, but it may cause abdominal pain, distension, or early satiety due to gross hepatomegaly (5, 8). More rarely, complications of cysts may include cyst infection (13), cyst hemorrhage (14), cyst rupture with hemoperitoneum (15), cyst torsion (14), portal hypertension (16), IVC or hepatic vein compression (17), or jaundice due to bile duct compression (18). We reviewed our experience with PLD at a tertiary care hospital to determine the natural history of this disease.

METHODS

Fifty-three patients with PLD were identified through retrospective chart review. Inpatient charts from 1990 to 2002

Table 1. Patient Characteristics (n = 53)

Gender (female/male)	33/20
Age at diagnosis (yr)	56.4 (SD 13.8, range 28–80)
Duration of disease at presentation (yr)	8.49 (SD 9.6, range 0–30)
Polycystic kidney disease	38 (71.7%)
Family history of polycystic liver disease	3 (5.7%)
Abdominal pain (n = 52)	19 (36.5%)
Dyspnea (n = 52)	5 (9.6%)
Restricted mobility (n = 52)	5 (9.6%)
Asymptomatic (n = 52)	33 (63.5%)
Hepatomegaly (n = 51)	23 (45.1%)
AST (IU/L)	27.2 (SD 27.6) normal range 0–40
Bilirubin (μmol/L)	14.6 (SD 20.7) normal range <20
Creatinine (μmol/L)	287.5 (SD 301) normal range 40–115
Alkaline phosphatase (IU/L)	142.2 (SD 205.9) normal range 30–130
Albumin (g/L)	35.0 (SD 6.5) normal range 35–50
5–10 hepatic cysts (n = 50)	2 (4%)
11–20 hepatic cysts (n = 50)	0 (0%)
>20 hepatic cysts (n = 50)	48 (96%)
Hepatic cyst diameter, maximal (cm)	7.73 (1–16)
Renal cysts present	47 (88.7%)
Renal cyst diameter, maximal (cm)	5.21 (1–11)
Hepatic cyst growth (n = 17)	6 (35.3%)

were identified by ICD 9 codes for PLD. Outpatient charts from a hepatology clinic (VB), a hepatobiliary surgery clinic (DB), and our liver transplant database were also reviewed to identify cases of PLD. Charts were reviewed for clinical information (gender, age, duration of disease prior to assessment, right upper quadrant pain, dyspnea, restricted movement, other symptoms, presence of autosomal dominant polycystic kidney disease, family history, hepatomegaly, treatments and response to therapy), laboratory information (AST, alkaline phosphatase, bilirubin, albumin, creatinine), and radiologic investigations (ultrasound or CT scan to determine the number and size of hepatic cysts, presence of complicated cysts, and presence of renal cysts). Where serial radiologic investigations were available, they were compared to assess cyst progression.

Each patient's family physician (as recorded in hospital records) was contacted to obtain current information regarding clinical, laboratory, and radiologic parameters.

Criteria for inclusion in this series were the presence of four or more hepatic cysts with no infectious etiology. This criterion has been shown to correlate with the genetic haplotype for PLD (9), and was subsequently shown to have a sensitivity of 81.8% and specificity of 100% in diagnosing PLD in patients over age 40 yr (8).

Descriptive statistics were derived using SPSS software (SPSS 11.0, Chicago, IL, USA). Ethics approval was obtained from the Health Research Ethics Board, University of Alberta Hospital, Edmonton, Canada.

RESULTS

Patient Demographics

Fifty-three cases of PLD were identified. Thirty-three patients were female (62.3%) and 20 patients were male (37.7%). The mean age at diagnosis of PLD was 56.4 yr (SD 13.8), with an 8.49-yr (SD 9.65) mean duration of disease prior to assessment at our center. A diagnosis of autosomal dominant polycystic kidney disease was found in 38 patients (71.7%). A family history of isolated PLD was recorded in three patients (5.7%) (Table 1).

Symptoms and Signs

Information regarding symptoms was available for 52 patients. Of these, 19 patients (36.5%) experienced abdominal pain. This was attributed to hepatomegaly in nine patients and cyst complications such as rupture, bleeding, or infection in two patients. Eight patients with pain had both hepatomegaly and cyst complications. Other symptoms were less common with dyspnea due to hepatomegaly in five patients (9.6%), and restricted mobility due to hepatomegaly in five patients (9.6%). Some patients reported more than one symptom, so that overall 33 patients (63.5%) were asymptomatic. Hepatomegaly was documented by the examiner in 23 of 51 patients (45.1%) with abdominal examination findings reported (Table 1).

Laboratory Data

Mean AST and bilirubin levels at the time of first evaluation were normal in this series, with values of 27.2 IU/L (range 4–173, normal range 0–40) and 14.6 μmol/L (range 5–126, normal range <20), respectively. The mean alkaline phosphatase value was elevated at 142.2 IU/L (range 41–1,183, normal range 30–130), as was the mean creatinine at 287.5 μmol/L (range 53–1,076, normal range 40–115), likely due to the concomitant polycystic renal disease seen in many patients. The mean albumin level was borderline reduced at 35.0 g/L (range 19–47, normal range 35–50) (Table 1).

Radiologic Imaging

All patients had evidence of multiple hepatic cysts consistent with PLD on either abdominal ultrasound or abdominal CT scan. A quantitative expression of the number of liver cysts was present in 50 patients. Of these, 2 patients (4%) had between 5 and 10 hepatic cysts, and 48 patients (96%) had more than 20 hepatic cysts. The mean diameter of the largest cyst in each patient was 7.73 cm (range 1–16 cm). Renal cysts were present in 47 patients (88.7%), with a mean maximal renal cyst diameter of 5.21 cm (range 1–11 cm). Seventeen patients had serial imaging over a mean follow-up time of 4.5 yr (range 0.5–13 yr), with cyst growth documented in six of these (35.3%) (Table 1).

Table 2. Clinical Outcomes (n = 40)

	Total Patients (n = 40)	With Renal Cysts (n = 28)	Without renal cysts (n = 12)
Follow up duration (yr)	4.69 (0.1–15)	5.09 (0.1–14)	3.75 (1–15)
Hepatic cyst bleeding	9 (22.5%)	8 (28.6%)	1 (8.3%)
Hepatic cyst rupture	5 (12.5%)	1 (3.6%)	4 (33.3%)
Hepatic cyst infection	5 (12.5%)	4 (14.3%)	1 (8.3%)
Invasive intervention	12 (30%)	8 (28.6%)	4 (33.3%)
Portal hypertension	1 (2.5%)	1 (3.6%)	0
Death	9 (22.5%)	7 (25%)	2 (16.7%)
Death attributable to PLD	1 (2.5%)	1 (3.6%)	0
Asymptomatic	26 (65%)	19 (67.9%)	7 (58.3%)
Persistent pain	3 (7.5%)	0	3 (25%)
Transplant	2 (5%)	2 (7.1%)	0

Clinical Outcomes

Follow-up information was obtained for 40 patients (75.5%) with a mean follow up duration of 4.69 yr (range 0.1–15 yr) (Table 2). Of this group, 9 patients experienced bleeding into a cyst (22.5%), 5 patients had a cyst rupture (12.5%), 5 patients developed an infected cyst (12.5%), 12 patients required a surgical procedure (30%) (Table 3), and 1 patient developed portal hypertension (2.5%). Nine patients died (22.5%), with death in one patient attributable to hepatic cysts. This was a 58-yr-old man, immunosuppressed for renal transplant due to polycystic renal disease. He died due to gram-negative sepsis with an infected hepatic cyst confirmed as the source. Other deaths were due to renal failure in one patient, pneumonia in two patients, gram-negative sepsis in one patient, lung cancer in one patient, intracranial hemorrhage in two patients, and a metastatic neuroendocrine tumor in one patient.

The clinical outcomes of patients with and without concomitant polycystic renal disease were compared. Although no formal statistical analysis was possible, there was a trend toward more bleeding into cysts and more cyst infections in patients with renal cystic disease. In addition, patients with renal cysts were more likely to experience death from any cause. Conversely, cyst rupture and persistent pain were more common in patients with isolated hepatic cysts. Of the 38 patients with polycystic renal disease, 23 required renal replacement therapy (dialysis or renal transplant). Patients requiring renal replacement were not more likely to develop a cyst complication than those without renal failure ($\chi^2 = 0.45$).

Of the patients requiring an invasive intervention, six were treated with cyst aspiration, three of whom had ethanol injection. One of the patients that underwent aspiration was the man who died of sepsis, although signs of sepsis preceded the aspiration. Relief of symptoms after aspiration was attained at least temporarily in the other five patients. Symptoms recurred in one patient after 3 months and in another after 1 yr. These patients were treated with repeat aspiration and surgical de-roofing with wedge resection, respectively. Symptom control was attained in the other two patients for 12 yr, and 2 months, respectively. A final patient died of pneumonia 1 yr after cyst aspiration.

Surgical de-roofing was performed in four patients. The patient who had de-roofing with wedge resection after failed cyst aspiration has been asymptomatic for 1 month. Another patient has been asymptomatic for 1 yr since de-roofing. Two others had recurrent symptoms at 2 yr and 5 yr following surgery, with the former patient having a repeat de-roofing procedure. Of the two patients who underwent liver resection, symptom-free status was attained for 4 yr in one, and 1 month in the other, who had concomitant cyst de-roofing. Two patients received a liver transplant (5%), one for portal hypertension and one due to markedly impaired quality of life from massive hepatomegaly. One transplant recipient developed posttransplant lymphoproliferative disorder during the first postoperative year, but with treatment is now in remission 2 yr posttransplant with excellent hepatic function and resolution of portal hypertension. She had previously received a renal transplant for polycystic renal disease. The other transplant recipient also experienced symptomatic relief, and has had good graft function for 4 yr posttransplant with mild renal impairment due to polycystic kidney disease. (Table 4)

At the last available follow up, 26 patients were asymptomatic (65%), 3 had persistent pain (7.5%), 2 had undergone transplantation (5%), and 9 had died (22.5%), although only one death was directly attributable to a complication of hepatic cystic disease.

Table 3. Invasive Interventions in Patients with Follow-Up (n = 40)

	Number of Interventions	Success Rate*
Aspiration	6 (15%)	3 of 6 (50%)
Ethanol ablation	3 (7.5%)	2 of 3 (67%)
De-roofing	4 (10%)	3 of 4 (75%)
Hepatic resection	2 (5%)	1 of 2 (50%)
Liver transplant	2 (5%)	2 of 2 (100%)

*Success defined as symptom resolution for 6 months or more.

DISCUSSION

The majority of patients in our series had multiple liver cysts in association with polycystic kidney disease (renal cysts in 88.7%). Hepatic cysts are the most common extra-renal manifestation of this disorder. Previous series have described age,

Table 4. Outcome of Invasive Interventions

Patient	Intervention	Outcome	Follow-up
Patient 1	Cyst aspiration	Death due to sepsis	—
Patient 2	Cyst aspiration	Symptom relief × 12 yr	Repeat aspiration 12 yr later
Patient 3	Cyst aspiration	Symptom relief × 1 yr	Died of pneumonia 1 yr later
Patient 4	Aspiration & EtOH	Symptoms recurred in 3 months	Repeat aspiration with symptom control × 6 yr
Patient 5	Aspiration & EtOH	Symptoms recurred at 1 yr	Surgical de-roofing with wedge resection with symptom relief at 1 month follow-up
Patient 6	Aspiration & EtOH	Symptoms recurred at 2 months	Repeat aspiration at 2 months with symptom control for 27 months
Patient 7	De-roofing	Symptom relief × 1 yr	No symptoms at 1 yr
Patient 8	De-roofing	Symptom relief	Gradual recurrence of symptoms by 5 yr
Patient 9	De-roofing	Symptom relief for 2 yr	Repeat de-roofing at 2 yr
Patient 10	Resection	Postop bile leak. Symptom relief × 4 yr	No symptoms at 4 yr
Patient 11	Liver transplant (portal hypertension)	PTLD 6 months posttransplant	2 yr posttransplant good graft function
Patient 12	Liver transplant (symptoms)	Good graft function	Graft function stable 4 yr posttransplant

female gender, parity, severity of renal cysts, and poor renal function as risk factors for the development of hepatic cysts in polycystic kidney disease (3–5). Our series supports these previous findings, as the majority of our patients were women, and both transplant recipients were female. The mean age at diagnosis was 56.4 yr, by which age previous studies have seen liver involvement in 40–56% of patients (4, 5). Furthermore, the mean creatinine in the group was more than twice the upper limit of normal. A trend toward a difference in disease outcome was also noted between patients with and without polycystic kidney disease. Patients with PKD were more likely to die, although only one death was attributable to hepatic cystic disease. Other causes of death included renal failure, infections, and a subarachnoid hemorrhage from an aneurysm. Thus, the increased mortality noted in patients with renal cysts is mostly due to other components of the PKD syndrome rather than the hepatic cysts. The higher rate of infected and bleeding cysts in patients with renal disease may be due to a more severe cyst burden, or to platelet or immune dysfunction in the setting of renal disease. The reason for a higher rate of cyst rupture and persistent pain in isolated PLD is more perplexing, although one could postulate a subcapsular location of liver cysts as a mechanism for pain and rupture.

Nonetheless, the majority of the patients in our series were asymptomatic (63.5%), which has been previously described (4). This is likely an underestimate of the true proportion of asymptomatic patients with PLD, since our series was composed solely of patients who had been referred to a tertiary care institution. Hepatic function was also well preserved despite the presence of massive hepatic cysts in many patients. The slight overall mean elevation in alkaline phosphatase may have been due to concomitant renal disease, but it has also been noted in isolated PLD and in the setting of acute hepatic cyst infection and may be related to biliary remodeling (8, 19). Preservation of hepatic function was seen despite severe hepatic cystic involvement with more than 20 cysts documented in 96% of patients. The proposed mechanism behind this finding is the preservation of normal hepatic parenchymal volume despite increasing cyst volume, as documented by CT scan studies (20).

A previous series of polycystic kidney disease patients on hemodialysis described a 10% mortality rate attributable to hepatic cyst complications (Table 5) (21). In contrast, only one death directly attributable to hepatic cysts was seen in our series. Thirty percent of patients did require a surgical intervention either due to symptoms or complicated cysts. Again, this high proportion may reflect referral bias to a tertiary care center.

Of the patients requiring surgery, the most enduring response was attained with liver transplantation or liver resection. Liver transplantation has been shown to have good symptomatic outcomes in these patients, but the benefits of subjecting a patient with normal hepatic function must be weighed carefully against the risks of surgery and immunosuppression (22). Cyst aspiration and fenestration procedures have been shown in other reports (23–26) to provide transient symptomatic relief, and in our study these procedures were also associated with the need for repeated intervention due to symptom recurrence.

Our study has several limitations due to the retrospective ascertainment of cases. We used the presence of four or more hepatic cysts as our inclusion criterion, which has been shown to be sensitive and specific for PLD in patients over age 40 yr (8). Using this cut-off could miss milder disease in younger patients who have not yet developed many cysts. Our results are also biased toward more severe disease due to referral bias, as most asymptomatic patients may not have been referred to a tertiary center. Follow up was incomplete mostly due to patients changing family doctors or moving. Again, the incomplete follow-up biases the results toward the more symptomatic end of the disease spectrum, as patients experiencing symptoms would be most likely to require follow up.

Table 5. Review of Case Series in the Literature

Author, Year	Number of Cases	Number with PKD	Mean Length of Follow Up	Number Symptomatic	Interventions
Van Erpecum *et al.*, 1987 (27)	15	7	4 yr	15* All had pain, Jaundice in 2 Ascites in 1	Aspiration in 4 Fenestration in 9
Eggink *et al.*, 1988 (28)	6	1	10.6 yr	2 Pain in 2	Fenestration in 1 Partial hepatectomy in 1
Ogutu *et al.*, 1991 (29)	14	14	None	14 All had pain or heaviness	None
Vauthey *et al.*, 1992 (30)	7	6	14 months	7* All had pain/pressure Dyspnea in 3 Early satiety in 3 Jaundice and ascites in 1, subsequent death due to sepsis Portal hypertension in 1 Acute abdomen from cyst bleeding in 1	Fenestration and resection in 5
Harris *et al.*, 1996 (31)	65	65**	Not specified	Pain in 12 Fullness in 3 Jaundice in 3 Ascites in 2 Death in 3 (due to abscess, sepsis, cholangitis)	Aspiration in 5 De-roofing in 3
Qian *et al.*, 2003 (8)	72	0	Not specified	38 Abdominal discomfort, dyspnea or early satiety 2 had cyst infections	21 Fenestration, partial hepatectomy (17) or liver transplant

*Series of highly symptomatic adult polycystic liver disease.
**Series of hepatic cystic disease in patients transplanted for PKD.

Encouragingly, despite the bias toward more severe disease, most patients experienced a benign disease course with pain as the most common symptom. Liver function generally remained normal and cyst complications were seen in only a minority of patients. The natural history of PLD was variable however, with some patients requiring invasive intervention or liver transplantation.

The summary of this experience would lead us to conclude that a majority of patients with PLD will experience a relatively benign and long clinical course. Few patients will succumb to the direct effects of PLD. Patients with advancing age and deteriorating renal function are most likely to experience complications of PLD. A conservative approach to treatment is therefore advocated with radiologic imaging and subsequent intervention guided by patient symptoms. Complicated hepatic cysts should be considered in the differential diagnosis of the patient with polycystic kidney disease who presents with either abdominal pain or fever. Minimally invasive interventions such as aspiration and sclerosis or fenestration are appropriate initial interventions, especially for patients with predominantly superficial cystic disease. Hepatic resection and liver transplantation are reserved for those with intractable and severe symptoms or life-threatening complications.

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