Dear Sir,

It is well known that cystic renal diseases (autosomal dominant polycystic kidney disease (ADPKD), autosomal recessive polycystic kidney disease (ARPKD) and nephronophthisis) can have hepatic alterations [1, 2]. Caroli’s disease has very rarely been described in ADPKD, ARPKD and nephronophthisis [3, 4]. There are two types of Caroli’s disease. In the diffuse type, cystic dilatation of the segmental bile ducts affects the whole intrahepatic biliary tree. In the localized type, cystic dilatation of the segmental bile ducts is confined to the left lobe. The former type is commoner than the latter and is usually associated with congenital hepatic fibrosis and portal hypertension. This diffuse type is transmitted as an autosomal recessive trait and may be associated with a renal malformation. The saccular intrahepatic bile ducts’ dilatations of Caroli’s disease can lead to cholangitis and gallstones.

We present the case of a 47-year-old patient without any family history of renal and hepatic diseases, with pathologic antecedents of acute pyelonephritis in her two pregnancies at the ages of 25 and 28. The first indications of renal failure appeared at the age of 38 (sCr 315 µmol/l). An abdominal computed tomographic scan showed a reduction in the size of the kidneys with small cortical cysts, small hepatic cysts, an asymptomatic gallstone and a cyst in the pancreatic head (fig. 1). A cystoureterography ruled out a vesicoureteral reflux.

At the age of 43 she developed a high fever of unknown origin. Antibiotic treatment was indicated and the fever ceased within 48 h. Then, at the age of 47, she had another bout of fever with discrete elevation of the aminotransferases. Blood cultures yielded Klebsiella pneumoniae; urine culture was negative. Acute cholecystitis was suspected after an ultrasonographic study, and a surgical operation was performed accordingly. A gallbladder stone was found and a whitened hepatic surface with diffuse fibrous stretches. A transcystic cholangiography per-reamputation showed a severe diffuse saccular dilatation of the intrahepatic bile ducts without obstruction (fig. 2). A cholecystectomy and hepatic biopsy was performed and Caroli’s disease was diagnosed. The current laboratory findings are: sCr 390 µmol/l (n = 44-106), ureas 22.6 mmol/l (n = 2.9-8.6), C1Cr 0.33 ml/s (n = 2), negative proteinuria, AST 16 U/l (n = 5-42), AST 18 U/l (n = 5-52), total bilirubin 8 µmol/l (n = 4-19), AP 97 U/l (n = 25-120), RBC 3.39 × 10^12/l (n = 4-5.2 × 10^12), Hct 31.4% (n = 38-45), Hb 10.4 g/dl (n = 12-16).
The diffuse form of Caroli’s disease is almost invariably associated with cystic lesions of the kidneys, mainly ARPKD and nephronophthisis; both usually manifested in childhood or adolescence. Association with ADPKD is extremely rare. Most of these cases are associated with hepatic fibrosis and portal hypertension. Our patient suffers from diffuse Caroli’s disease with an associated cystic renal disease but not hepatic fibrosis or portal hypertension. The kidney disease does not fulfil criteria of nephronophthisis (the good evolution of renal function in relation to the patient’s age, the grade of anemia and the cortical location of the renal cysts), nor criteria of ARPKD or ADPKD. We believe it is due to the possible genetic heterogeneity of these diseases and their low incidence [5].

To summarize, in patients with polycystic renal disease associated with liver cysts who develop fever of unknown origin, Caroli’s disease should be suspected and the appropriate tests should be carried out to confirm the diagnosis.

Fig. 1. Abdominal computed tomographic scan shows a reduction in the size of the kidneys, renal cysts, small hepatic cysts, and a cyst on the head of the pancreas.

Fig. 2. Transcystic cholangiography shows marked saccular dilatations of the intrahepatic bile ducts without obstruction.

References
Caroli’s Disease and Cystic Kidney Disease Nephron 1996;73:310-311.