

# Polycystic disease: a rare indication for combined liver and kidney transplantation

AKK Chui, D Koorey, OP Pathania, ARN Rao, GW McCaughan, AGR Sheil

**We report on a 52-year-old woman who presented with polycystic disease. Both of her kidneys had been removed and she had undergone one failed kidney transplantation. She had severe symptoms from the polycystic liver. The diseased liver and kidney were both treated successfully by performing a combined liver and kidney transplantation.**

*HKMJ 2000;6:116-8*

*Key words: Cysts/surgery; Kidney, polycystic; Kidney transplantation; Liver diseases; Liver transplantation*

## Introduction

Adult polycystic liver disease (APLD) is a rare, benign, and dominantly-inherited disorder characterised by multiple cystic lesions throughout the liver. The prevalence of APLD from an autopsy study has been shown to range from 0.13% to 0.6%.<sup>1</sup> Between 75% and 90% of patients with APLD have associated renal disease. The majority of patients with APLD remain free of liver-related symptoms. In some patients, however, the presence of symptoms due to cyst enlargement and compression of nearby structures necessitates surgical intervention. Several interventions, such as cyst aspiration with or without sclerotherapy, cyst fenestration, and liver resection, have been reported as treatments of symptomatic APLD.<sup>2</sup> Single-donor combined liver and kidney transplantation for polycystic disease has been performed so far in 14 patients, as reported in the English literature.<sup>1,3-5</sup> This is the first Australasian report of combined liver and kidney transplantation for polycystic liver and kidney disease.

## Case report

A 56-year-old woman presented to the Royal Prince Alfred Hospital in July 1995 with polycystic kidney disease. The disease had been diagnosed in 1960,

when she had presented with renal failure, for which haemodialysis was commenced. She had undergone bilateral nephrectomy because of haematuria and loin pain in 1982. By 1984, her liver had enlarged considerably and she had acquired hepatitis C, although the route of transmission was uncertain. She had received a renal transplant in 1992, but in February 1995, a graft nephrectomy had been performed because of chronic rejection of the graft. Haemodialysis was then recommenced.

At the time of presentation to the Royal Prince Alfred Hospital, the patient had tender hepatomegaly. Her appetite was poor, she had muscle wastage, and massive refractory ascites had developed. Despite the largeness of the liver and the massive ascites, she weighed only 55 kg. Laparoscopic marsupialization and fenestration of the hepatic cysts were performed on two occasions but provided little relief. A liver biopsy confirmed chronic active hepatitis C. Computed tomography showed that the inferior vena cava and the portal vein were compressed by the huge polycystic liver (Fig 1). The hepatic venous wedge pressure was 30 mm Hg and the caval pressure was 10 mm Hg, thus confirming portal hypertension. The following results from serum biochemical analysis demonstrated an impairment of liver function: total bilirubin, 10 mmol/L (normal range, 2-18  $\mu$ mol/L); albumin 22 g/L (40-60 g/L); alkaline phosphatase, 132 U/L (30-120 U/L); alanine aminotransferase, 92 U/L (0-35 U/L);  $\gamma$ -glutamyltransferase, 82 U/L (0-30 U/L); aspartate aminotransferase; 26 U/L (0-36 U/L); international normalised ratio (INR), 1.1. Because of the progressive symptoms and complications of polycystic disease, combined liver and kidney transplantation was considered.

**Royal Prince Alfred Hospital, Camperdown NSW 2050, Australia:  
Australian National Liver Transplantation Unit**

AKK Chui, MB, BS, FRCS (Edin)

OP Pathania, MB, BS, MS

ARN Rao, MB, BS, MS

AGR Sheil, MA, FRCS

**AW Morrow Gastroenterology and Liver Centre**

D Koorey, FRACP, PhD

GW McCaughan, FRACP, PhD

**Correspondence to: Dr AKK Chui**



Fig 1. Computed tomography scan of the liver showing diffuse cysts and the compression of the inferior vena cava

On 27 September 1995, orthotopic liver transplantation followed by renal transplantation were performed using liver and kidney transplants from a 61-year-old man. The intra-operative blood loss was approximately 12 L. The liver and renal grafts functioned well immediately after transplantation. Postoperatively, the patient required ventilatory support for 1 day in the Intensive Care Unit. Immunosuppressive therapy consisted of prednisolone, cyclosporin A, and azathioprine. The mass of the resected liver was 2.9 kg after cyst evacuation (Fig 2). Histological examination of the resected liver showed typical signs of APLD and confirmed chronic active hepatitis C. The patient experienced one mild episode of liver graft rejection, which responded to an increased prednisolone dosage. She was discharged home 53 days after the operation. During the 2-year follow-up period, results of renal function tests remained

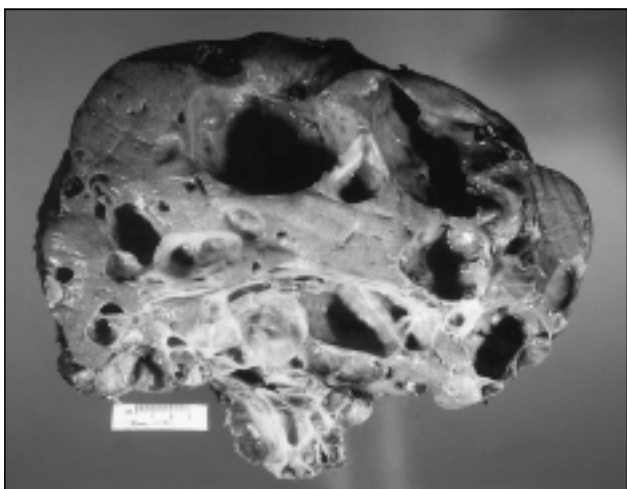


Fig 2. Resected specimen of the polycystic liver

within the normal ranges and she has enjoyed good health. The results of the liver function tests were as follows: alkaline phosphatase, 158 U/L; alanine aminotransferase, 125 U/L;  $\gamma$ -glutamyltransferase, 294 U/L; aspartate aminotransferase, 73 U/L. These results indicated a recurrence of hepatitis C, but the synthetic function of the graft was shown to be good (bilirubin, 12  $\mu$ mol/L; albumin, 46 g/L; INR, 1.0).

## Discussion

The availability of haemodialysis and kidney transplantation has meant that patients with polycystic disease, who previously would have died of renal failure, are now surviving longer. Consequently, there have been more reports<sup>2,6</sup> of patients with polycystic disease, in whom symptoms and/or complications develop because of hepatic cysts. Taking conservative non-surgical approaches to treat patients with severely symptomatic APLD has limited value.<sup>6,7</sup> While the transhepatic fenestration procedure described by Lin et al<sup>8</sup> decreases cyst tension and relieves abdominal discomfort, this procedure does not permanently reduce liver size, and hence long-term relief cannot be expected. Liver resection, with or without transhepatic fenestration, is a more popular technique and achieves favourable results in some patients with massive APLD. This technique, however, has been associated with a prolonged postoperative recovery period and significant morbidity, especially in cases with associated chronic renal failure.<sup>6</sup> Such surgical procedures also result in the development of adhesions, thus making subsequent liver transplantation more difficult.<sup>9</sup>

In addition, the management of secondary complications of portal hypertension in patients with APLD can prove difficult. Peritoneovenous shunting is generally ineffective in providing lasting benefit for refractory ascites.<sup>10</sup> Portal-systemic shunting causes significant surgical mortality and morbidity and is unlikely to be beneficial, as in this case, when the inferior vena cava is compressed by the liver. Transjugular intrahepatic portal-systemic shunting is difficult to perform in patients with APLD, because both the hepatic and portal veins can often be compressed by cysts. In this case, chronic active hepatitis with impaired liver function precluded the possibility of successful liver resection. The symptoms and complications of APLD had progressed to the stage where the patient's quality of life was poor and the continued survival threatened. Performing combined liver and kidney transplantation was thus appropriate, and the procedure proved successful.

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