

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

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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.

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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%.¹ 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.² Single-donor combined liver and kidney transplantation for polycystic disease has been performed so far in 14 patients, as reported in the English literature.^{1,3-5} 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 μ 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); γ -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.

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References

1. Kwok MK, Lewin KJ. Massive hepatomegaly in adult polycystic liver disease. *Am J Surg Pathol* 1988;12:321-4.
2. Sanchez H, Gagner M, Rossi RL, et al. Surgical management of non-parasitic cystic liver disease. *Am J Surg* 1991;161:113-9.
3. Taylor JE, Caine RY, Stewart WK. Massive cystic hepatomegaly in a female patient with polycystic kidney disease treated by combined hepatic and renal transplantation. *QJM* 1991;293:771-5.
4. Klintmalm GB, Busuttill RW. Combined liver-kidney transplantation. In: Klintmalm GB, Busuttill RW, editors. *Transplantation of the liver*. Philadelphia: WB Saunders; 1996:514-7.
5. Flye MW, Brian FD, Donna LP, Lloyd ER, Mohankumar T. Protective effects of liver transplantation in simultaneously transplanted kidney in a highly sensitized patient. *Transplantation* 1990;50:1051-4.
6. Van Erpecum KJ, Janssens AR, Terpstra JL, Tjon A, Thom RT. Highly symptomatic adult polycystic disease of the liver. *J Hepatol* 1987;5:109-17.
7. Saini S, Mueller PR, Ferrucci JT, Simeone JF, Wittenberg J, Butch RJ. Percutaneous aspiration of hepatic cysts does not provide definite therapy. *AJR* 1983;141:559-60.
8. Lin TY, Chan CC, Wang SM. Treatment of non-parasitic cystic disease of the liver: a new approach to therapy with polycystic liver. *Ann Surg* 1968;168:921-7.
9. Henne-Bruns D, Klomp HJ, Kremer B. Non-parasitic liver cysts and polycystic liver disease: results of surgical treatment. *Hepatogastroenterology* 1993;40:1-5.
10. Elcherth J, Vons C, Franco D. Role of surgical therapy in management of intractable ascites. *World J Surg* 1994;18:240-5.



ROYAL AUSTRALASIAN COLLEGE OF SURGEONS 2000 ANNUAL SCIENTIFIC CONGRESS

MELBOURNE, 7-12 MAY 2000

The Melbourne Annual Scientific Congress is to be held at the Melbourne Exhibition and Convention Centre complex from 7-12 May 2000.

Features of the meeting will include the Ecumenical Service to be held at St. Paul's Cathedral on Sunday 7 May 2000. This will be followed by the Convocation in the evening at the Melbourne Exhibition and Convention Centre at which the Former Governor General of Australia, the Right Honourable Sir Ninian Stephen, will be the Syme Orator.

On both Monday 8 May 2000 and Tuesday 9 May 2000 there will be a number of combined sessions with the Australian and New Zealand College of Anaesthetists who are meeting at the Crown Towers Hotel across the Yarra.

The Social Programme includes Sectional Dinners on Tuesday 9 May 2000 and Thursday 11 May 2000 and the Congress Formal Dinner on the evening of Wednesday 10 May 2000. On the evening of Monday 8 May 2000 the RACS will be hosting an evening in the lively Southgate precinct which will include street theatre groups, wine tasting and dining.

- A copy of the Provisional Programme and Registration Form can be found via the College website at the following address: www.racs.edu.au
- Registration enquires can be directed to the ASC Manager, Mr Kevin Wickham, Telephone +61 3 9859 6899 or Email: kevin@wickhams.com.au
- General enquiries can be directed to the ASC Secretariat, Ms Kate Young, Telephone +61 3 9249 1200 or Email: kate.young@racs.edu.au