Renal replacement therapy in a patient with phocomelia

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The drug thalidomide, launched in 1954 as an antiemetic for use in pregnancy, was responsible for an epidemic of birth deformities (limb phocomelia) that affected some 2500 children worldwide, about 400 of these in the UK. When this condition is complicated by end-stage renal failure, treatment presents exceptional difficulties.

CASE HISTORY

Born in 1962 with four-limb phocomelia due to maternal use of thalidomide, the patient was diagnosed during her second pregnancy in 1992 as having adult polycystic kidney disease (APKD). At that time she was working as a computer programmer, but she became a full-time mother in 1994. She was first referred to renal transplant surgeons at the age of 37 (late 1999) with creatinine 250 $\mu\text{mol/L}$ and glomerular filtration rate 20 mL/min. She enjoyed a remarkably independent life with her two children and a live-in carer. Dialysis was obviously going to be very difficult. Her rudimentary limb buds would not support any form of vascular access and peritoneal dialysis was considered impractical because of her lack of arms and also the presence of two large polycystic kidneys. The best option would have been a planned living-related transplant before dialysis was needed, but this was impossible because of the family history of APKD and the unsuitability of her father. The only alternative was to place her on the waiting list for a cadaveric renal transplant when she became symptomatic, in the hope of avoiding the need for long-term haemodialysis via a central venous dialysis line. Although clinically there were no pulses in the groins, magnetic resonance imaging and computer tomography indicated the presence of a normal aorta, inferior vena cava, and common iliac arteries and veins. Despite the presence of two large polycystic kidneys, there was enough space in the iliac fossae to allow renal transplantation.

Her renal function slowly deteriorated. Special consideration was given to her case by UK Transplant and they allowed her to be placed on the clinically urgent waiting list in March 2001. She subsequently received a cadaveric renal transplant in May that year. A routine extraperitoneal approach to the left iliac fossa was made. The external iliac vessels were vestigial but the common and internal iliac arteries and veins were normal. A right kidney with single vessels was transplanted, with the renal artery and vein anastomosed end-to-side onto the common iliac vessels. The HLA mismatch was 2-0-1 and she received ciclosporin, mycophenolate mofetil and prednisolone. The donor was cytomegalovirus (CMV) positive and recipient negative so she also received valaciclovir prophylaxis. The potential difficulties with venous access for blood taking and drug administration were avoided by perioperative placement of a right internal jugular Hickman line.

She had good primary function and was discharged on day 12 with a creatinine of 75 $\mu\text{mol/L}$. Protocol biopsies on days 7 and 28 showed minimal changes with no evidence of acute rejection. Her subsequent progress was marred by active CMV disease requiring treatment with intravenous ganciclovir via the Hickman line and a bilateral native nephrectomy for recurrent infections and bleeding into cysts. However, 2 years after her transplant she remains well with a serum creatinine of <100 $\mu\text{mol/L}$.

COMMENT

This patient suffered the double blow of an inherited autosomal dominant renal disease and a severe drug-induced birth deformity. Despite the daunting prospect of surgery, a combination of good planning, extensive preoperative imaging, flexibility in organ allocation and a team approach succeeded in maintaining a good-quality and independent life.

Increasingly, patients who would once have been judged unsuitable are presenting as potential candidates for transplantation. Examples are those with complex lower urinary tract abnormalities or multiple earlier transplants onto the iliac vessels. In many of these patients, long-term access to the central veins or the peritoneum has been exhausted, and in these circumstances renal transplantation is similar to both thoracic and liver transplantation in its ability to save the life of the individual patient. This case demonstrates that difficult patients need not be excluded from a renal transplant programme. However, it is not always easy to balance the needs of the individual patient, whose potential for a favourable outcome will be well below average, against the use of a precious resource such as a donated cadaveric kidney.

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