Dear Sir,

The use of hydroxyurea to increase HbF production in patients with homozygous sickle cell disease is increasing, particularly after recent publication of data demonstrating a significant reduction in painful crises [1]. We here describe the successful use of hydroxyurea in a sickle cell patient with severe painful crises occurring after renal transplantation and an associated rise in haematocrit.

A 26-year-old Jamaican with homozygous sickle cell disease had a stable haemoglobin of 8-9 g/dl and painful crises once every 3 years. He developed proteinuria and renal impairment, and renal biopsy demonstrated mesangiocapillary glomerulonephritis. Renal function slowly declined with a parallel fall in haematocrit and he commenced haemodialysis in early 1994. At this point his haemoglobin was 4 g/dl but he failed to respond to subcutaneous erythropoietin despite doses of up to 600 U/kg/week – higher doses were not tolerated because of pain on injection and the usual causes of erythropoietin resistance were excluded.

In December 1994 he received a cadaveric renal allograft after a 4-unit exchange transfusion, with standard triple immuno-suppression (cyclosporin 8 mg/kg b.d. initially, prednisolone 20 mg and azathioprine 1 mg/kg). Three further units of packed red blood cells were given peri-operatively. Creatinine fell to 159µmol/l (1.8mg/dl) and haemoglobin rose to 11 g/dl with HbS > 80% once the transfused blood had gone. The e-c-120-1
14 100
300 400
Time (days)

Fig. 1. Haemoglobin concentration and globin fractions in relation to renal transplantation and hydroxyurea therapy.

Renal transplantation in homozygous sickle cell patients with end-stage renal disease is well described with good graft and patient survival rates [2, 3]. The usual rise in haematocrit, seen when the graft functions well, is sometimes associated with the development of painful crises, ascribed to the concurrent rise in plasma viscosity [4, 5]. Previously these crises have been managed with analgesia and venesection or exchange trans-
vated haemoglobin heralded the onset of severe painful sickle crises requiring prolonged hospital admission for analgesia and venesection. Three months after transplantation he commenced hydroxyurea 500 mg b.d. (17 mg/kg/day) with rapid and complete cessation of painful episodes. Haemoglobin has fallen to around 10 g/dl and the hydroxyurea dose has been titrated to maintain this level and to allow for mild leucopenia.

References

Hydroxyurea, Sickle Cell Disease, Renal Transplantation
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