Prevalence of autosomal-dominant polycystic kidney disease in Alentejo, Portugal

To the Editor: In a recent issue of *Kidney International*, Levy and Feingold reviewed studies estimating the prevalence of single-gene progressive kidney diseases [1]. We would also like to report an ongoing study on the prevalence of autosomal-dominant polycystic kidney disease (ADPKD) in Alentejo, in the south of Portugal, where a different rate was found. Preliminary data were reported at the EDTA Congress held in September 1999, in Madrid, Spain (abstract; de Almeida et al, *Nephrol Dial Transplant* 14:A101, 1999). Patients and their families were tracked from dialysis centers and nephrology outpatient services and all nephrologists in the region participated. By the prevalence day, July 31, 1999, we identified 84 patients with ADPKD, 16 with renal cysts but with no family history of renal cysts, and 160 first-degree relatives with a 50% risk; therefore, a total of 180 affected persons were expected. According to the last census (1991), Alentejo has 543,442 inhabitants, which gave a prevalence rate of 1:3019. Calculating prevalence rates of monogenic traits based on a priori risk is expected to overestimate the true prevalence of the disease. On the other hand, underdiagnosis and follow-up by non-nephrologists often underestimates the prevalence. A more accurate prevalence is expected when this study is finished and all general practitioners will report on their patients at that time.

In summary, we present a lower prevalence for ADPKD than reported by Levy and Feingold [1], a result more consistent with those of Davies et al [2] in the Welsh population. Whether this number is the result of a similar method or whether it truly represents the prevalence of ADPKD in Alentejo remains to be established.

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Post-transplant recurrence of focal segmental glomerulosclerosis

To the Editor: We read with interest the article by Sharma et al, describing the blocking effect of normal sera on the focal segmental glomerulosclerosis (FSGS) activity [1]. Their experiments showed that preincubation with normal serum followed by washing and subsequent incubation with FSGS serum resulted in the protection in permeability effect of FSGS factor(s).

Posttransplant recurrence of FSGS has been reported in 30–50% of patients and the outcome is miserable. To prevent this recurrence we have used plasmapheresis preoperatively with satisfactory results. The methods and results have been reported elsewhere [2, 3]. In brief, pretransplant plasmapheresis is repeated 2 to 3 times immediately before transplantation and in each session 50 to 75 mL/kg of the patient's plasma is exchanged with albumin-Ringer's solution or fresh-frozen plasma. In our study, the incidence of recurrence in children with prophylactic plasmapheresis was reduced by one half, compared to those who received no prophylaxis. Furthermore, even when recurrence occurred, the degree of proteinuria was markedly less severe.

Focal segmental glomerulosclerosis appears to be the immediate consequence of a primary or secondary glomerular epithelial cell defect or injury. In fact, typical FSGS lesions appeared later in such patients, as with persistent proteinuria initiated immediately after transplantation in our series. Therefore, the first injury by the so-called FSGS factor(s) may lead to further glomerulosclerotic lesions. We speculate that pretransplant plasmapheresis in our trial may be effective not only in reducing FSGS factor(s), but also in blocking the effect of FSGS serum with normal serum transfused as substitution fluid.

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