Sir, 

During long-term dialysis, the kidney is at maximum risk for the development of renal cell carcinoma (RCC), and the occurrence of bilateral cases is not rare [1,2]. Based on previous studies, bilateral nephrectomy is no longer favoured, primarily due to increased morbidity and mortality after the management and complications of the anephric state, namely anaemia, hypocalcaemia and hypotension from dysadrenalism [3,4].

Between October 2004 and September 2005, we performed bilateral synchronous nephrectomy by a transperitoneal approach for suspected cases of RCC and spared one or both of the adrenal glands in seven patients. The patients with long-term haemodialysis periods ranging from 6.5 to 24.5 years (mean ± SD 16.2 ± 6.3) included four cases of acquired cystic disease of the kidney and three cases of autosomal dominant polycystic kidney disease in which kidney size was >20 cm. The mean kidney weight was 1677 g (mean ± SD 1677 ± 1420), mean operative time was 288 min (mean ± SD 288 ± 66) and mean estimated blood loss was 1045 ml (mean ± SD 1045 ± 66). During the convalescent period, there were no mortalities. During the follow-up period of 6 to 37 months, haematocrit, serum calcium and aldosterone were maintained from 27.5 to 34.1% (mean ± SD 32.0 ± 2.2), 9.8 to 11.6 mg/dl (mean ± SD 10.7 ± 0.8) and 2.3 to 22.3 pg/ml (mean ± SD 12.3 ± 8.3), respectively. Genetic recombinant erythropoietin derivatives were administered to six cases, and vitamin D was administered to one case after the operation. Hypotension (systolic blood pressure <100 mmHg during haemodialysis) from dysadrenalism was not noted at the end of the follow-up period. Out of the 14 specimens, there were 10 RCC, 2 oncocytoma and 4 benign complex cysts. Lung metastasis occurred in one patient, while the other six patients were asymptomatic and had no tumour recurrence.

The results demonstrated that bilateral synchronous open nephrectomy is practical with acceptable morbidity, due to advances in medical management and surgical techniques. In the review of literature, respectable en bloc renal size on bilateral synchronous laparoscopic nephrectomy ranged from 20 to 27 cm. However, such cases can have a risk of dissemination, due to cyst disruption during circumferential mobilization and extraction of the kidney [5,6]. In order to avoid these complications, we selected long-term dialysis patients with a kidney size >20 cm. Based on the kidney size and the clinical condition, further studies are needed in order to determine the indication of bilateral synchronous open or laparoscopic nephrectomy for bilateral RCCs following long-term dialysis.

Conflict of interest statement. None declared.

Advanced Access publication 23 December 2007

Bilateral synchronous open nephrectomy for renal tumours in patients following long-term dialysis

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doi: 10.1093/ndtplus/sfm033

Advanced Access publication 23 December 2007

Organ transplantation law in Pakistan to curb kidney trade: chance for global reflection

Sir,

Statistics on the number of kidney transplantations by individual facilities (particularly origins of the donors and recipients) are not disseminated in Pakistan, one of the major ‘Kidney Outlets’, where a minimum of 4000 unethical kidney allo-transplantations take place annually, accounting for 20% of illegal allografts worldwide [1]. The absence of a transplant registry is highlighted by marked variance in approximations for 2006 (Table 1). Concealment of data on Karachi is noticeable [2].

Fifty to seventy-five percent of the recipients are foreigners [3]. The annual turnover for this trade is around US $20 million. Effectively, we share the disease burden of overseas communities [4]. This translates into a dialysis:transplant ratio of 10:1 for the local population.

Efforts to establish ethical transplant practices in Pakistan date from 1988. Following the intervention by the Supreme Court of Pakistan in July 2006, a law to regulate organ transplantation (and curbing the burgeoning kidney trade in particular) was drafted by the law ministry, in January 2007. This draft had to be revised in August 2007, in order to accommodate reservations on the part of the judiciary on the clauses dealing with compensation for donors. Transplantation of Human Organs and Tissues Ordinance 2007 stands promulgated as of 3 September 2007. A related bill awaits ratification by lawmakers.

This bill is based on promotion of living-related donations, according to the ministry of health. A closely matched
non-related person may only provide an organ out of compassion when a genetically-related donor is unavailable. Cadaveric donation is not among the designated priorities. The edict imposes strict penalties for brokers and those indulging in unsupervised removal, storage and grafting of organs. A federal monitoring authority and evaluation committees in designated institutions have been convened. A donor from their country of origin will have to accompany non-residents if they want to avail of our services [5].

Transplant registry, a nationwide pool of potential donors, a donor card system, as well as allocation of resources for disadvantaged end-stage renal disease (ESRD) patients, donor care and follow-up are planned. Transplant institutions will only be allowed to function if they (and personnel thereof) meet prescribed licensure criteria. Limited experience (28 cadaveric grafts to date), non-existence of a brain-death law and doubts about public approval of cadaveric mode of organ harvesting preclude the optimistic notion of the Transplant Society of Pakistan that a deceased organ donation programme will help save 50 000 lives every year.

Pakistani law has global implications because of rampant transplant tourism [6]. It is consistent with contemporary trends [7]. Organ donation by relatives should be portrayed as an act of dignity. I suggest that a sustained strategy of directed donations be adopted, with more effective early identification of a genetically-related donor for each ESRD patient. A strong family setup, and motivation by specialist counsellors, may both play a critical role.

Conflict of interest statement. None declared.

### Table 1. Approximations of renal transplantations in Pakistan for 2006

| Source                      | GoP\(^a\) | WHO       | Transplant Society of Pakistan | Newspapers [3] | Foreign journal\(^b\) | The Transplant Society |
|-----------------------------|-----------|-----------|-------------------------------|----------------|-----------------------|------------------------|
| Quoted figure              | 1600–2200 | 600–700   | 3000–4500                     | 3000           | 4000\(^b\)            | 2000                   |

\(^a\)Government of Pakistan.
\(^b\)India Journal of Medical Ethics [1].

Sir,

Fabry disease is a rare X-linked lysosomal storage disorder caused by mutations in the \(\alpha\)-galactosidase \(A\) (\(\alpha\)-gal A) gene. Affected hemizygous males have impaired or abrogated activity of \(\alpha\)-gal A enzyme that leads to a buildup of neutral glycosphingolipids, particularly globotriaosylceramide (Gb3), in many tissues and organs [1]. Symptoms include acroparesthesias, angiokeratomas, corneal opacities, and cardiovascular and renal complications, with death usually occurring by the fifth decade of life [1]. Females that are heterozygous carriers of an \(\alpha\)-gal A enzyme mutation show a wide variety in the range and severity of their symptoms.

The kidney is of particular pathological importance in Fabry disease, as kidney failure due to Gb3 buildup is a major complication; along with cardiac failure it is a leading cause of death [2]. In recent years, it has been proposed that some dialysis patients could be suffering from undiagnosed Fabry disease and therefore there would be benefits to screening dialysis patients for this disease [3,4]. We carried out a pilot screening study at the Toronto General Hospital in Ontario, Canada, to implement our previously published high-throughput blood spot assay for Fabry disease [5]. Informed consent was obtained from 147 forthcoming dialysis patients to screen their blood for levels of \(\alpha\)-gal A enzyme. Based on our blood spot assay, 141 of these patients had whole blood \(\alpha\)-gal A enzyme activity levels within the normal range compared to normal and Fabry controls [5, and unpublished data]. For those six patients with activity below 1.5 nmol/h/ml (<65% of average enzyme activity in normal controls), secondary plasma \(\alpha\)-gal A enzyme assays showed that all patients were within the normal range.

While no Fabry patients were identified in the relatively small participating patient population, this study shows application of our previously published high-throughput screening method. Our assay is a rapid and low-cost method for screening patients that may be suffering from undiagnosed Fabry disease. This initial screening study did not exclude any patients willing to participate, so both males and females were included. As with other symptoms, \(\alpha\)-gal A enzyme levels in female Fabry patients vary with the individual and can even approach normal levels. We thus cannot discount the possibility that there remain undiagnosed