Correspondence

Risk factors have special relevance for living kidney donors

Sir,
Awareness of modifiable risk factors for end-stage renal disease (ESRD)\(^1\,^2\) is likely to be of great benefit to living kidney donors, so as to reduce the risk of complications such as hypertension and diabetes, which hasten the onset of ESRD.\(^1\,^2\) The risk of hypertension can be mitigated through lifestyle counselling, which includes dietary advice, weight management and regular exercise,\(^3\) and the same is true of the risk of type 2 diabetes.\(^4\,^5\,^6\,^7\) Given the fact that, at a mean of 12.2 years (Standard Deviation 9.2 years) after kidney donation, 32.1\% of living donors have hypertension, and 12.7\% have albuminuria,\(^8\) identification of hypertension before and after kidney donation is crucial to prognosis because, in one study, both hypertension and proteinuria were identified as risk factors for ESRD in living kidney donors, notwithstanding the suboptimal follow-up of living kidney donors in that study.\(^9\) For optimum identification of hypertension in prospective donors, ambulatory blood pressure monitoring should complement office blood pressure measurements so as to identify masked hypertension,\(^10\) and the same logic should apply to long-term postoperative surveillance of donors. In the event of an increase in blood pressure, what is arguable is whether treatment should be initiated when blood pressure is in the range 130/80–140/90 mmHg or whether the conventional threshold of 140/90 mmHg should apply. In either event, it would seem reasonable to set the ‘goal’ blood pressure at the <130/80 mmHg level applicable to patients with chronic renal disease,\(^11\) and to diabetics,\(^12\) so as to reduce the risk of further deterioration in renal function in a manner analogous to the reduction in the risk of deterioration in renal function documented in previous studies involving patients with chronic renal failure and diabetes, respectively.\(^11\,^12\) In the event of co-existence of left ventricular hypertrophy, a further refinement would be to include, in the treatment goals, reduction of blood pressure already in the normal range so as to achieve further regression of left ventricular mass,\(^13\) given the fact that ‘combined cardiac and renal involvement defines a very high-risk phenotype affected by worse prognosis than any single damage’.\(^14\) In short, such is our indebtedness to living kidney donors (including literary icons such as Adrian Mole)\(^15\) that, even a single preventable donation-related fatality in that population is one death too many.

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References

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A 33-year-old gentleman presented in December 2002 with complaints of giddiness, vomiting, abdominal pain, loose motions and pigmentation around the umbilicus. He had confirmed hyponatremia (Na-122 mEq/l) and a subnormal synacthen test (cortisol at 30 min was 287 nmol/l) and was diagnosed with Addison’s disease and was commenced on hydrocortisone and fludrocortisone replacement. He had a stable disease throughout except for two episodes of Addisonian crisis secondary to gastroenteritis requiring intravenous steroids. Six years later, he presented with complaints of bladder disturbance (polyuria, urgency and dysuria). He was hospitalized thrice in a very short span of time with confirmed urinary tract infections. One such episode led a complete retention of urine for which he had to be catherized for 2 days. Digital examinations of the prostate, ultrasound of the urinary tract and flow metric studies were all normal. Investigations revealed normal glucose, potassium, calcium, plasma and urinary osmolality. The only abnormality noted then was presence of patchy alopecia and absence of hair on lower limbs and axillae. A year later, he started to develop leg symptoms in the form of weakness and progressively worsening mobility. He had difficulty in protracted standing and walking but still managed to do things for himself independently. This was not accompanied by any sensory, bowel or bladder symptoms and clinical examination of his lower limbs was quite unremarkable.

We tried to explore his recent neurological presentation by revisiting his family history, which was very interesting (Figure 1). His half brother presented at the age of 7 years initially (1978) with an inability to write followed by an unsteadiness of gait and standing and walking but still managed to do things for himself independently. This was not accompanied by any sensory, bowel or bladder symptoms and clinical examination of his lower limbs was quite unremarkable. Six years later, he presented with complaints of bladder disturbance (polyuria, urgency and dysuria). He was hospitalized thrice in a very short span of time with confirmed urinary tract infections. One such episode led a complete retention of urine for which he had to be catherized for 2 days. Digital examinations of the prostate, ultrasound of the urinary tract and flow metric studies were all normal. Investigations revealed normal glucose, potassium, calcium, plasma and urinary osmolality. The only abnormality noted then was presence of patchy alopecia and absence of hair on lower limbs and axillae. A year later, he started to develop leg symptoms in the form of weakness and progressively worsening mobility. He had difficulty in protracted standing and walking but still managed to do things for himself independently. This was not accompanied by any sensory, bowel or bladder symptoms and clinical examination of his lower limbs was quite unremarkable.

We tried to explore his recent neurological presentation by revisiting his family history, which was very interesting (Figure 1). His half brother presented at the age of 7 years initially (1978) with an inability to write followed by an unsteadiness of gait. Subsequently, he developed weakness of arms and legs, incoordination of limbs, incontinence and myoclonic jerks. His developmental milestones were normal but had clinical evidence of mental retardation, spasticity of arms and legs, facial and tongue muscle weakness with equinus deformity of his feet. He was noted to have bilateral optic atrophy on opthalmoscopy and his brain scan showed a large fourth ventricle and low attenuation changes in the white matter. An electroencephalogram revealed gross abnormal activity of the posterior half of brain and an electromyogram revealed generalized reduction in action potential and amplitude. He was tested for enzymes of metabolic pathways, all of which were within normal limits. His autoantibodies for thyroid, parietal cell, smooth muscle, mitochondria, adrenal and pituitary were also within normal limits. Urine was negative for

**Importance of family history in patients with adrenoleukodystrophy**

Sir,

X-linked adrenoleukodystrophy accounts for up to 10% cases of adrenocortical insufficiency and is now being increasingly identified in young males with idiopathic Addison’s disease. Widespread pathological damage is encountered in the nervous system due to abnormal accumulation of very long chain fatty acids (VLCFAs). The disease often demonstrates intrafamilial phenotypic variability, suggesting that non-genetic factors play a role in such expression. A detailed history and early estimation of VLCFAs in clinically suspected individuals make diagnosis possible in affected families. We present an interesting case of adrenoleukodystrophy where diagnosis was helped by a significant family history and also a brief review of the contemporary literature.

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