haemodialysis showed leucocytes 24 090/mm³ (94.2% neutrophils; 0% eosinophils), haemoglobin 11.0 g/dl and platelets 202 000/mm³. Blood glucose was 222 mg/dl, sodium 143 mEq/l, potassium 3.8 mEq/l, calcium 9.6 mg/l, albumin 3.4 g/dl, uric acid 6.6 mg/dl, amylase 75 U/L, lipase 65 U/L, alanine aminotransferase (ALT) 30 U/l, aspartate aminotransferase (AST) 46 U/l, creatinine kinase-MB (CK-MB) 10.9 U/l, troponin-I 0.13 ng/ml, C-reactive protein quantitative 0.2 mg/dl and IgE 344.4IU/ml. Blood gas analysis showed pH 7.47, PaO₂ 80.1 mmHg, O₂ saturation 96.1%, and PCO₂ 35 mmHg. Echocardiograms revealed adequate left ventricular function with mild mitral valve regurgitation. During the follow-up, regular haemodialysis was given three times a week with cellulose diacetate hollow fibre (Dicea170G, Baxter, USA), and no anaphylaxis occurred.

In summary, we describe a delayed near-fatal anaphylactic reaction induced by a polysulphone haemodialyser. Although hypersensitivity to biocompatible dialysers is rare and usually occurs during initial haemodialysis, doctors and members of haemodialysis centers should keep in mind that a risk of severe anaphylaxis may occur at any time during a prolonged haemodialysis treatment.

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Haemodialysis in a patient with haemophilia B

Sir,

I would like to raise a problem of preparation for haemodialysis (HD) in patients with a mild type of haemophilia B. Chronic renal failure is a very rare condition in haemophiliacs and/or individuals with other hereditary clotting disorders and the dialysis modality for these patients may be a difficult choice.

A 53-year-old man with haemophilia B, mild hypertension, hepatitis C virus infection (anti-HCV positive) developed end-stage renal disease (ESRD), most probably due to chronic glomerulonephritis. Preparation for HD was undertaken in accordance with the patient’s choice (HD or PD) at an eGFR of 12 ml/min/1.73 m². Laboratory data showed a prolonged activated partial thromboplastin time (aPTT) of 82 s and factor IX activity of 8.6% (mild type of haemophilia B). All other coagulation factor levels were normal. It is worthy of note that the patient was admitted to our clinic with a diagnosis of severe haemophilia B, based on a factor IX activity of 0.1%, assessed 24 years previously.

To create the vascular access, intensive treatment with coagulation factor IX was given to achieve the target level (35–40%) for small surgical procedures. The calculated dose was administered, as recommended, intravenously 1 h before creation of the fistula and then every 12 h for two consecutive days. The successfully created wrist arteriovenous fistula was complicated by a small haematoma. Twenty days after fistula creation, the patient started HD using a small-size (17 G) single needle without systemic/local anticoagulation. Two-needle dialysis was introduced 4 weeks after fistula placement. During the 6 months of follow-up, no bleeding episodes after needle removal and no clotting in dialyzers or drains were observed. There was no need for factor IX substitution during and after the dialysis sessions.

The modality of dialysis in patients with hereditary clotting disorders is a difficult choice. When considering haemodialysis, intensive treatment with coagulation factors is crucial for vascular access creation and in some severe cases for the HD procedure as well. Peritoneal dialysis has been performed infrequently in haemophiliacs. The clinical results of PD treatment may fluctuate between an uncomplicated course, when coagulation replacement therapy is required only during peritoneal catheter placement, and rather serious complications such as haemoperitoneum episodes [1].

An important issue in the preparation for renal replacement therapy is the evaluation of factor IX activity. In our patient we noticed an increase in factor IX activity within 24 years (from 0.1 to 8.6%), which resulted in a lower dose of substituted factor IX prior to the surgical procedure. In recent publications, factor IX, a circulating serine protease, has been shown to increase with age in humans [2]. Kurachi et al. [3] investigated the mechanisms of the age regulation of the human factor IX gene. They identified two genetic elements: an age-related stability element (ASE, GAGGAAG) and an age-related increase element (AIE, a unique stretch of dinucleotide repeats), which were responsible for age-related stable and increasing expression patterns, respectively. The molecular weight of factor IX has been estimated to be between 65 and 72 kDa, depending on the method of assessment [4]. Therefore it is unlikely that a higher level of factor IX is because of low GFR (cutoff of the glomerular basement membrane ~60 kDa) [5].

In conclusion, HD is a good therapeutic option for patients with a mild type of haemophilia B and ESRD requiring dialysis. Prolonged aPTT allows for HD without anticoagulation. Re-evaluation of the severity of haemophilia is advisable, due to the age-related increase in factor IX activity which may contribute to the proper management of the patient.

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**Post-transplant focal segmental glomerulosclerosis refractory to plasmapheresis and rituximab therapy**

Sir,

Primary focal segmental glomerulosclerosis (FSGS) recurs in 20 to 50% of kidney transplants [1]. Recently, there have been reports demonstrating the successful use of rituximab for recurrent post-transplant FSGS [2–4]. We report a case of recurrent FSGS that was refractory to both plasmapheresis and rituximab therapy.

A 48-year-old female with biopsy-proven FSGS underwent living kidney transplantation. She received anti-thymocyte globulin, tacrolimus, mycophenolate mofetil and prednisone. Her immediate post-transplant course was uneventful and her serum creatinine was 1.0 mg/dl on day 2. Fifteen days post-transplant, she was edema free, her proteinuria had fallen from 7.8 to 3.4 g/day and she was not on an ACE-inhibitor or angiotensin receptor blocker. One month post-transplant, she had edema, a weight gain of 4.7 kg, and her serum creatinine was 1.0 mg/dl on day 2. Fifteen days post-transplant, she was edema free, her proteinuria had fallen from 7.8 to 3.4 g/day and she was not on an ACE-inhibitor or angiotensin receptor blocker.

A transplant biopsy was performed, which showed no evidence of tubulitis or segmental sclerosis (cortico-medullary junction not sampled). Immunofluorescence showed 1+ IgM staining in the mesangium and C4d was negative. Electron microscopy showed diffuse effacement of the foot processes and there were no electron dense deposits. The pathological findings were consistent with recurrent FSGS [1]. Plasmapheresis was started and she received 13 exchanges over 4 weeks. There was no response; a second renal biopsy was thus performed, which showed ongoing evidence of FSGS. Plasmapheresis was restarted and she received another 13 exchanges over 4 weeks. After the final exchange, she had 6.41 g/day of proteinuria. Due to the lack of response, she was given six doses of rituximab (600 mg or 375 mg/m² per dose) over 8 weeks. Unfortunately, there was no response and she had 5.42 g/day of proteinuria.

Rituximab, a monoclonal antibody directed against CD20, has been reported by several investigators to successfully treat post-transplantation FSGS [2–5]. The first two reports describing rituximab for recurrent FSGS were unique, in that they both occurred in the setting of post-transplant lymphoproliferative disorder [2,3], while the subsequent two cases were not [5]. It is not clear why our patient was resistant to rituximab therapy, given the reported experience in the literature to date. It is unlikely to be related to insufficient dose, as she received a total of 3600 mg of rituximab and her B cells were depleted with therapy (0.1% CD19 and 0.2% CD20 positive B cells 4 weeks post-rituximab).

In conclusion, we have shown that rituximab, given in an adequate dose, does not always lead to a remission in recurrent FSGS. These findings suggest that equipoise exists regarding the use of rituximab for post-transplant FSGS and that randomized trials are needed to properly address this issue.

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