

## Case Report

# Renal replacement therapy in a patient with hemophilia A: a case report

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We describe the case of a male hypertensive patient with severe hemophilia A. In August 1999, he was admitted to our nephrology department, with hemarthrosis, severe hypertension, dyspnea with minimal efforts, increasing blood urea nitrogen, anemia, uremic symptoms, reduced urine volume, mild edema of lower limbs and no hyperkalemia. Image exams confirmed the diagnosis of End Stage Renal Disease. A Tenckhoff peritoneal dialysis catheter was inserted. He then started continuous ambulatory peritoneal dialysis. In August 2005 he evolved with peritoneal failure (peritoneal equilibration test showing ultra-filtration disorder-I), and was transferred to hemodialysis. A permanent catheter insertion into the right subclavian vein was scheduled. Hemodialysis sessions lasted 4 hours, three times a week, and gradually resulted in hemodynamic stabilization. In September 2005, an arteriovenous fistula was placed in the right forearm between the cephalic vein and the radial artery. In January 2007, the patient was admitted with abdominal and epigastric pain, double lumen catheter infection, peritoneal catheter infection, globoid tympanic abdomen and mild pain on palpation. Propaedeutics showed a large preperitoneal hematoma with bowel compression. Due to the catheter infection, we decided to puncture the fistula using a 17G needle. Apart from some bleeding during and after the beginning of hemodialysis, there were no other fistula complications. The patient evolved with progressive worsening of clinical symptoms, died in February 2007. In summary, an individualized treatment plan, mainly regarding adequate hemostatic monitoring, care of the dialysis access, multiprofessional and family involvement, may contribute to a successful management of hemophilia patients undergoing dialysis.

**Keywords:** Chronic kidney failure; hemophilia A; renal dialysis

## INTRODUCTION

Hemodialysis in hemophiliacs with chronic renal failure is

almost unknown in literature.

The hemophilia is a hereditary disease, linked to X chromosome, caused by deficiency of factor VIII, causing disturbance in the mechanism of blood coagulation (Rossi-Ferreira et al., 2006). It is manifested by the

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susceptibility of the patient have episodes of spontaneous bleeding or post-trauma (Bellegard and Caetano, 2006).

Although, hematuria is a common manifestation of abnormal hemostasis in hemophiliacs, the chronic renal failure is unusual in these cases (Small et al., 1982).

In hemophiliacs, the hemodialysis sessions generally become more worrisome in function of the problems of change in the normal clotting process.

The following report refers to a patient with hemophilia who required dialysis, which made use of factor VIII concentrate as a strategy to control the coagulopathy.

## CASE REPORT

Chronic renal failure is an uncommon disease in patients with hemophilia and other coagulation disorders and hemodialysis may be a difficult choice for such individuals (Bajo et al., 2000; Kuzstal et al., 2008). The following report refers to the first and only hemophiliac patient treated in our unit, with a history of End Stage Renal Disease (ESRD) undergoing renal replacement therapy.

We describe the case of a male hypertensive patient with severe hemophilia A (residual Factor VIII activity <1%) who was born in 1957 and had hematological follow-up since childhood at the Hemominas Foundation of Belo Horizonte, Minas Gerais, Brazil. In his medical history there were repeated episodes of hemarthrosis, bleeding gums and delay in homeostasis of small vessels. He was a carrier of both hepatitis C and B viruses (anti-HCV positive and HBsAg positive) with no signs of active hepatitis.

In August 1999, he was admitted to our nephrology department, with hemarthrosis, severe hypertension, dyspnea with minimal efforts, increasing blood urea nitrogen (serum creatinina: 19.5 mg/dl, urea: 246 mg/dl), anemia (Hb: 9.3 g/dl), uremic symptoms, reduced urine volume (700 ml/24h), mild edema of lower limbs and no hyperkalemia ( $K^+$ : 4.2 mmol/l). Since a renal biopsy was not possible, image exams confirmed the diagnosis of ESRD, most likely due to glomerulonephritis secondary to hepatitis virus infection. A Tenckhoff peritoneal dialysis catheter was inserted under local anesthesia, with replacement of Factor VIII concentrate one hour before the procedure. There were no major complications, only a mild to moderate bleeding at the site of catheter insertion. He evolved with a voluminous abdominal and pelvic hematoma which responded to replacement of Factor VIII. After six days of catheter implantation and no evidence of bleeding, Factor VIII was suspended and indicated only in case of bleeding.

After 25 days, the patient was discharged from hospital. He then started continuous ambulatory peritoneal dialysis (CAPD) for six years.

During this period, he had two episodes of bacterial peritonitis which were treated with cephalothin and gentamicin. In March 2005, he had lung congestion, anasarca and severe arterial hypertension. The peritoneal equilibration test (PET) was requested for CAPD monitoring at 0, 2 and 4 hours (result showing ultra-filtration disorder-III). We decided to change dialysis bags more frequently (seven times a day), and to substitute CAPD for continuous automated ambulatory peritoneal dialysis (CAAPD). In August 2005 he evolved with peritoneal failure (PET showing ultra-filtration disorder-I), and was transferred to hemodialysis.

A permanent catheter insertion into the right subclavian vein was scheduled. Laboratory data showed prothrombin activity 59% and partial thromboplastin time 64" (27"). The procedure had no complications and was performed with replacement of Factor VIII. Bleeding was observed at the site of catheter insertion during nine days. For the treatment we positioned the patient's head at a 45 degree angle, we used compression dressings and replaced Factor VIII. Hemodialysis sessions lasted 4 hours, three times a week, and gradually resulted in hemodynamic stabilization. Heparin was suspended (5,000 IU of heparin circulating in the system during 5 minutes, priming discarded and saline flushes every 30 minutes during hemodialysis). Catheter anticoagulation consisted of tract filling with dilute heparin solution (1000 IU in each tract).

In September 2005, an arteriovenous fistula was placed in the right forearm between the cephalic vein and the radial artery (3500 IU Factor VIII one hour before surgery and 2500 IU every twelve hours for three days after the procedure). There were no complications during surgery and no postoperative bleeding.

In January 2007, the patient was admitted with abdominal and epigastric pain, double lumen catheter infection (up to date there had been three more episodes of infection), peritoneal catheter infection, globoid tympanic abdomen and mild pain on palpation. Propaedeutics showed a large preperitoneal hematoma with bowel compression (3500ml estimated volume). Due to the catheter infection, we decided to puncture the fistula using a 17G needle. Apart from some bleeding during and after the beginning of hemodialysis, there were no other fistula complications.

The patient evolved with progressive worsening of clinical symptoms, severe pericardial effusion with diastolic compression of the right chamber. He was confused, sedated, feeding through a nasogastric open tube, with bilious drainage and, later on, parenteral nutrition. He eventually died in February 2007.

## DISCUSSION

The motivation to write it came from noticing a lack in the literature on the theme, when we sought for orientation on whether or not to dialyse a hemophilic patient, and which would be the best dialysis form to use in this case. Although it is rare to have hemophilic patients in need for dialyses treatment, clinical management of hemophilia associated to renal failure needs to be discussed by the international scientific community for deliberation on best procedures in these situations.

We are confident that sharing our experience will contribute to further reflection on clinical management of dialytic hemophilic patients, helping the establishment of adequate and efficient strategies to treat these patients.

Dialysis treatment in hemophiliacs when indicated can be accomplished with the administration of factor VIII concentrate and of platelets. The need for any intervention depends on the stage of disease, coagulation status and assessment of the hematologist, who can determine the best therapy to be adopted in accordance with the needs of each patient. We opted for prophylactic replacement therapy of factor VIII concentrate due to the good results in the literature with using this therapeutic modality.

In summary, an individualized treatment plan, mainly regarding adequate hemostatic monitoring, care of the dialysis access, multiprofessional and family involvement, may contribute to a successful management of hemophilia patients undergoing dialysis.

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