was started 16 months ago, the patient has not required any hospital admission, performs a normal physical activity, and has substantially recovered renal function. Hemodialysis discontinuation is not considered appropriate.

While it is true that our patients could be considered in some case potentially recoverable, considering the severity of the baseline condition and/or underlying disease, no statement could be made a priori. Special mention should be made of the improved quality of life and absence of hospital admissions once replacement therapy was started. The indication for monitoring of residual function is emphasized.[9,10]

1. Sekkarie MA, Port FK, Wolfe RA, Guire K et al. Recovery from end-stage renal disease. *Am J Kidney Dis* 1990; 15: 61-5.
2. Levy JB, Hammad T, Coulthart A, Dougan T, Pusey CD. Clinical features and outcome of patients with both ANCA and anti-GBM antibodies. *Kidney Int* 2004; 66 (4): 1535-40.
3. Jenette JC. Rapidly progressive crescentic glomerulonephritis. *Kidney Int* 2003; 63 (3): 1164-77.
4. Serra A, Romero R. Fracaso renal agudo en las vasculitis sistémicas asociadas a anticuerpos anticitoplasma de los neutrófilos (ANCA) en pacientes de edad avanzada. *Nefrología* 2001; 21: 1-8.
5. Knudsen LM, Hjoth M, Hippe E. Renal failure in multiple myeloma: reversibility and impact on the prognosis. Nordic Myeloma Study Group. *Eur J Haematol* 2000; 65 (3): 175-81.
6. Bladé J, Fernández-Llama P, Bosch F, Montolíu J et al. Renal failure in multiple myeloma: presenting features and predictors of outcome in 94 patients froma a single institution. *Arch Intern Med* 1998; 158 (17): 1889-93.
7. Marenzi G, Grazi M, Assanelli, Campodonico J et al. Circulatory respons to fluid overloada renoval by extracorporeal ultrafiltration in refractory congestive Heart failure. *J Am Coll Cardiol* 2001; 38 (4): 963-8.
8. Sheppard R, Panyon K, Pohwani AL, Kapoor A et al. Intermittent outpatient ultrafiltration for the treatment of sever refractory congestive heart failure. *J Card Fail* 2004; 10 (5): 380-3.
9. Gámez C, Teruel JL, Ortuño J. Evolución de la función renal residual en enfermos tratados con hemodiálisis. *Nefrología* 1992; 12: 125-9.
10. Rodríguez Benítez P, Gómez Campderá FJ, Rengel M, Anaya F. Recuperación de la función renal en pacientes en programa de diálisis. *Nefrología* 2002; 22 (1): 92-3.

L. Quiñones Ortiz, A. Suárez Laurés, A. Pobes Martínez and R. Forascepi Roza
Servicio de Nefrología. Hospital Cabueñes. Gijón.
Correspondence: *Luis Quiñones Ortiz. luysquio@hotmail.com. Hospital Cabueñes. Camino de los Prados, 395. 33203 Gijón (Asturias). España.*

Recovery of total innmunoglobulin and immunoglobulin subclasses in nephrotic syndrome: deflazacort *vs* methylprednisone

Nefrología 2008; 28 **(5)** 563

To the editor: Hypogammaglobulinemia in nephrotic syndrome (NS) is a condition attributable to increased catabolism and urinary loss of immunoglobulins.[1,2]

Patients with NS have an increased risk of infection, primarily caused by capsulated germs (pneumococci, *Hemophilus influenzae).*[3,4] There is evidence suggesting that IgG2 are antibodies protecting against pneumococci.

Our purpose was to study recovery of the different immunoglobulin subclasses in patients with NS treated with methylprednisone and deflazacort during remission and relapse.

PATIENTS AND METHODS

Eleven patients with a mean age of 48 months (range, 16-52 months) were studied. An interventional, single blind, clinical study, randomized for treatment start and with treatment crossover after the first relapse, was designed.

Methylprednisone (MPD) was given at 48 mg/m^2/day for 6 weeks, followed by 2/3 of the dose every other day for the next 6 weeks.

The equivalent dose of deflazacort (DFZ) was 72 mg/m^2/day (maximum 90 mg/day), with the same therapeutic scheme.

Blood samples were taken at relapse and 40 days after remission was achieved to measure total IgG and its subclasses by the radial immunodiffusion method.

RESULTS

Mean times on remission were 7.8 ± 0.36 days with MPD and 8.3 ± 0.22 days with DFZ.

Mean times to relapse were 85 ± 3.8 days with MPD and 102 ± 4.19 days with DFZ.

Both total IgG and IgG1 similarly recovered with both corticosteroids, but IgG2 and IgG3 only significantly increased with DFZ. Percent recovery of IgG and its subclasses was asymmetric during remission.

IgG1 showed the greatest recovery, while IgG2 reached 50% of normal value. The same imbalance was found with both treatments.

While this was not considered as an objective, it may be stated that the incidence rate of infection by capsulated germs was not significant with both corticosteroids (p 0.12) and that less adverse effects occurred when DFZ was used (hypertrichosis, Cushingoid appearance, ocular hypertension, hyperglycemia).

CONCLUSIONS

We found an imbalance in recovery of IgG subclasses during remission of NS treated with both corticosteroids, but a better recovery of such levels was achieved with DFZ.

1. Longsworth IG, MacInnes DA. An electrophoretic study of nephrotic sera and urine. *J Exp Med* 1998; 71: 77-82.
2. Oxelius V-A. IgG Subclass levels in infancy and childhood. *Act Pediatr Scand* 1979; 68: 23-27.
3. Lowy Y, Brezin C, Neauport-Sautes C, Theze J, Fridman WH. Isotype regulation of antibody production. T-cell hybrids can be selectively induced to produce IgG1 and IgG2 subclass-specific suppressive immunoglobulin-binding factors. *Proc Natl Acad Sci USA* 2003; 80 (8): 2323-7.
4. Lowy Y, Theze J. Regulation of IgG1 and IgG2 subclass expression by adjuvant-activated splenic suppresser T-cell Cell. *Immunol* 1999; 91 (2):467-76.

M. Liern, S. Diéguez and C. Canepa
Servicio de Nefrología. Hospital de Niños Ricardo Gutiérrez. Buenos Aires. Argentina.
Correspondence: *José Miguel Liern. jliern@yahoo.com. Hospital General de Niños Ricardo Gutiérrez. Alberti, 1880. 1714 Ituzaingo (Buenos Aires). Argentina.*

Rhabdomyolysis after correction of severe hyponatremia due to an attack of acute intermittent porphyria

Nefrología 2008; 28 **(5)** 563-564

To the editor: A vascular mechanism responsible for renal damage during at-

tacks of acute intermittent porphyria (AIP), due to sustained vasospasm, appears to exist.[1] Hyponatremia is a common complication in this disease, that is usually related to a syndrome of inappropriate secretion of antidiuretic hormone (SIADH).[2,3] Rhabdomyolysis may be a frequent complication during hyponatremia correction in which elevation of creatine phosphokinase (CPK) levels runs parallel to the recovery of sodium levels and renal function is not usually impaired.

We report the case of a 16-year-old female patient born in Mauritania, with a family history of acute intermittent porphyria, who was admitted to hospital for abdominal pain and vomiting for the past several days associated to an impaired consciousness level and two episodes of generalized tonic-clonic seizures, after which she remained in a postictal state and required orotracheal intubation and mechanical ventilation. Laboratory test results include severe hyponatremia (Na 95 mM/L; normal, 135-145) associated to plasma (244 mOsm/L) and urinary hyposmolarity (222 mOsm/L), urinary frequency and data suggesting hypovolemia (central venous pressure of 4 mmHg). Isotonic saline infusion was therefore started. Rhabdomyolysis was detected 24 hours after admission. Maximum CPK levels of 35628 U/L were found at 48 hours, but no renal function impairment was noted. Patient clinical signs and history suggested an AIP attack, that was confirmed by measuring in a spot urine sample levels of delta-aminolevulinic acid of 39.7 mg/g (normal, 0-5), as well as values in 24-hour urine of 154.3 mg of porphobilinogen (normal, 0-2), 484 µg/24 h of coproporphyrin (normal, 0-60), and 1,471 µg/24 h of uroporphyrin (normal, 0-22). Once diagnosis was confirmed, treatment was started with intravenous human hemin at 3 mg/kg/day for 4 days. The patient developed arterial hypertension that required administration of beta-blockers. Clinical course was satisfactory, with a progressive improvement in consciousness level. Mechanical ventilation was withdrawn 6 days after admission, and patient was discharged with a normal consciousness level and no symptoms. A genetic study to search for mutations and screen for heterozygous family carriers was requested.

AIP is the most common and severe of hepatic porphyrias.[4] Disease transmission is autosomal dominant, and the gene encoding AIP is located in the long arm of chromosome 11. AIP causes deficiency of the enzyme uroporphyrinogen I synthetase, formerly called porphobilinogen synthetase (PBG), thereby blocking heme synthesis. The most typical clinical signs include nausea, vomiting, constipation, diarrhea, urinary retention, tachycardia, hypertension, mental symptoms, and muscle pain and weakness. These attacks may be triggered by barbiturates, anticonvulsants, estrogens, oral contraceptives, alcohol, or low-calorie diets. Seizures may occur in 20% of cases, particularly in patients with hyponatremia.

The relationship between hyponatremia and rhabdomyolysis may apparently result from intracellular potassium efflux to compensate for the cell edema caused by decreased sodium levels, which would cause a decrease in transmembrane potential and hence in muscle metabolism.[5] Other authors postulate that it is hyponatremia correction itself which causes changes in muscle cells ion concentrations and osmolarity resulting in incapacity to maintain homeostasis in cell volume regulation. This would cause membrane fragility and muscle enzyme release into the bloodstream.[6]

1. Anderson C, Wikberg A, Stegmayr B, Lithner F. Renal symptomatology in patients with acute intermittent porphyria. A population-based study. *J Intern Med* 2000; 248 (4): 319-325.
2. Salgueira M, Monroy A, Jarava C, Molas R, Páez MC, Amor J et al. Hiponatremia en la porfiria aguda intermitente. Un problema diagnóstico en urgencias. *Nefrología* 1997; 17 (1): 88-91.
3. López Montes A, Lorenzo I, Pérez Martínez J. Síndrome de secreción inadecuada de hormona antidiurética y porfiria. *Nefrología* 2004; 24 (Extr 3): 85-88.
4. Anderson KE, Bloomer JR, Bonkovsky HL, Kushner JP, Pierach CA, Pimstone NR, Desnick RJ. Recommendations for the diagnosis and treatment of the acute porphyrias. *Ann Intern Med* 2005; 142 (6): 439-50.
5. Trimarchi H, González J, Olivero J. Hyponatremia-associated rhabdomyolysis. *Nephron* 1999; 82 (3): 274-7.
6. Rizzieri DA. Rhabdomyolysis after correction of hyponatremia due to psychogenic polydipsia. *Mayo Clin Proc* 1995; 70 (5): 473-6.

M. García-Martul, L. Santana-Cabrera, Z. Santos-Moyano and M. Sánchez-Palacios

Servicio de Medicina Intensiva. Hospital Universitario Insular de Gran Canaria.

Correspondence: *M. García-Martul. mgarciamartul@yahoo.es. Hospital Universitario Insular de Gran Canaria.*

Biliptysis as the initial symptom of a rare complication of autosomal dominant polycystic kidney disease

Nefrología 2008; 28 **(5)** 564-565

To the editor: Autosomal dominant polycystic kidney disease (PKD) is an inherited disease usually occurring in adults and characterized by gradual development of multiple renal and extrarenal cysts during life, with different and unpredictable growth rates.[1] This disease occurs in approximately 1 out of every 1,000 people, and is the most common polycystic kidney disease.

This letter discusses the case of a 62-year-old female patient, a Caucasian with chronic renal failure (CRF) secondary to PKD who attended the office complaining of fever for the past 4 days, followed by right chest pain irradiating to hypochondrium and right flank, associated to cough and breathlessness. Some days later she suffered increased breathlessness and cough, which produced an intense yellow expectoration, was sudden, and left an unpleasant taste (biliptysis). The patient experienced right pleural effusion. Minimum undifferentiated pleurotomy was performed, and broad spectrum antimicrobial treatment was administered. She subsequently showed an image with the appearance of a walled abscess in the right base, in addition to pneumothorax, which prompted the decision to perform conventional surgery, at which a biliobronchial fistula from the lower pulmonary lobe and a hepatic cyst, joined together and fibrosed with the phrenic nerve in the fistulous tract area, were found (fig. 1).