

Results: 3 patients with initial resistance to steroids showed response to intravenous pulse cyclophosphamide after 4 injections. 4 patients with initial resistance to steroids developed CRF during the observation period. The other eighteen patients with late steroid resistance responded to intravenous pulse cyclophosphamide, but all were steroid dependent at the end of the observation period. Five could not be weaned from steroids during the intravenous pulse cyclophosphamide treatment period. The other thirteen patients achieved relatively prolonged remission (7 months to 36 months).

Conclusion: Not all patients with initial steroid resistance respond to intravenous pulse cyclophosphamide. The side effects of intravenous pulse cyclophosphamide were negligible. Beneficial therapy for initial steroid-resistant FSGS has been remaining important.

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PLASMAPHERESIS IN TREATMENT OF PEDIATRIC PATIENTS WITH NEUROLOGICAL DISEASES: 10 YEARS EXPERIENCE

S. Rödl¹, I. Marschitz², C.J. Mache³, U. Gruber-Sedlmayr⁴, B. Plecko⁴, G. Zobel¹

¹Department of Pediatrics, PICU, Medical University Graz, Graz, ²Dept. of Paediatrics, Neonatology, Paracelsus Medical University Salzburg, Salzburg, ³Department of Paediatrics, Nephrology Unit, ⁴Department of Paediatrics, Neurology Unit, Medical University Graz, Graz, Austria

Objective: The aim of our study was to determine the outcome of children with severe life threatening symptoms treated with plasmapheresis (PF).

The goal of PF is to either remove autoantibodies / toxins or substitution of a missing protein by fresh frozen plasma (FFP) replacement.

Methods: Setting: 12 bed interdisciplinary paediatric ICU;

Patients: 15 patients; 4 male, 11 female; mean age: 13 years, treatment sessions with PF: 1 - 11.

Indications for PF by diagnoses with severe organ failure: Systemic Lupus Erythematoses (SLE) 4 pts, Myasthenia Gravis(MG) 5 pts, chronic inflammatory demyelinating polyneuropathy 2 pts, Good Pasture

Syndrome (GPS) 1pt, Chorea minor 2 pt, acute demyelinating encephalomyelitis (ADEM) 1 pt;

Additional immunosuppression (10 of 11): steroids (PRED), Cyclosporine A (CsA), mycophenolate mofetil (MMF), cyclophosphamide (CYC) and rituximab (RTX).

Results: The 4 patients with SLE and the patient with GPS with neurological symptoms improved within the first 2 weeks of PF treatment and recovered completely. Additional immunosuppression was necessary by nephrological indication. Two patients with SLE were transplanted successfully.

Our 5 patients with severe MG had complete remission with additional immunosuppression for more than 3 years. The 2 patients with chorea minor improved during PF and recovered within 6 weeks. The patients with CIDP and ADEM had both residual symptoms.

Conclusion: In paediatric patients with severe organ failure resulting from a diagnosis indicating plasmapheresis, the combined treatment with plasmaphereses and immunosuppression may lead to persistent remission of organ failure.

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RESCUE TREATMENT WITH HEMODIAFILTRATION IN INFANTS WITH END-STAGE RENAL FAILURE AFTER COMPLICATIONS ON PERITONEAL DIALYSES

S. Rödl¹, I. Marschitz², C.J. Mache³, M. Koestenberger⁴, G. Madler¹, T. Rehak¹, G. Zobel¹

¹Department of Pediatrics, PICU, Medical University Graz, Graz, ²Dept. of Paediatrics, Neonatology, Paracelsus Medical University Salzburg, Salzburg, ³Department of Paediatrics, Nephrology Unit, ⁴Department of Pediatrics, Cardiology Unit, Medical University Graz, Graz, Austria

Objective: Mortality in infants with end-stage renal disease (ESRD) during early life is still high. The aim is the presentation of the efficacy of hemodiafiltration (HDF) in infants with ESRD and complications on peritoneal dialysis (PD).

Patients and Methods: We show the alternative treatment with HDF in 4 infants requiring renal replacement therapy for ESRD before 28 days of life at a tertiary University Hospital from 1999 to 2010. Diagnoses leading to ESRD were cortical

necrosis after perinatal shock, renal dysplasia in prune belly syndrome, neonatal Shigatoxin-associated hemolytic uremic syndrome, and bilateral nephrectomy in autosomal recessive polycystic kidney disease.

Results and Discussion: Initially, 3/4 patients were started with HDF. Reasons were abdominal surgery in two patients and presumed acute renal failure in one patient. Thereafter all patients were switched to PD. If complications occurred on PD, patients were switched back to HDF. In case of severe volume overload we started with cHDF and continued with iHDF. During the first year of life, 3/4 patients required 1 to 3 HDF rescue treatment periods (major abdominal surgery in 5 periods, recurrent peritonitis in 1 period). 1/4 patients had to be switched to maintenance iHDF due to multiple abdominal leaks at 8 months of age. All 4 patients survived the first year of life. One patient is on maintenance PD at 23 months. Two patients were transplanted successfully at 22 and 35 months, respectively.

In conclusion, in case of complications on PD, HDF is an option to achieve long-term survival and kidney transplantation.

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ONE YEAR CLINICAL EXPERIENCE WITH THE PRISMAFLEX HF 20 SET IN FOUR INFANTS

S. Rödl¹, I. Marschitz², C.J. Mache³,
M. Koestenberger⁴, G. Madler¹, T. Rehak¹,
G. Zobel¹

¹Department of Pediatrics, PICU, Medical University Graz, Graz, ²Dept. of Paediatrics, Neonatology, Paracelsus Medical University Salzburg, Salzburg, ³Department of Paediatrics, Nephrology Unit, ⁴Department of Pediatrics, Cardiology Unit, Medical University Graz, Graz, Austria

Objective: We present the first clinical report involving use of the Prismaflex (Gambro) with the newly developed Prismaflex HF20 Set in 200 consecutive treatments for patients with end-stage renal disease (ESRD).

Patients: Four infants with ESRD (ages of 13, 10, 5, and 25 months, and with a body weight of 9.5 kg, 8.5, 6.5, and 9.5 kg, respectively) were treated with intermittent hemodiafiltration (HDF). All patients received five hour treatments. Treatment monitoring included weight change and fluid balance of

patients, the efficacy of treatment, the number of interventions, and machine alarms.

Results: We used blood flow rate 50 ml/min, dialysate fluid rate of 1000 ml/h and replacement fluid rates of 500 ml/h. Based on the precision of the Prismaflex scales, desired fluid balance was achieved according to the prescribed weight loss for the patients. The efficacy of treatment was monitored by BUN and creatinine values (55 ± 20 mg/dl and 5.3 ± 1.0 mg/dl, respectively) at start of treatment and their decrease after 4 hours of RRT (21 ± 8 mg/dl and 2.2 ± 0.6 mg/dl, respectively). At the start of treatment, urea and creatinine clearances were 23.3 ± 7.0 ml/min and 19.1 ± 4.4 ml/min, respectively. No complications during the HDF occurred.

Conclusion: This is the first report about the use of the Prismaflex HF20 Set in infants with ESRD. No side effects were detected, treatments were tolerated well in all patients, and adaptation of the flow rates to the infants' needs was good.

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NEPHROLITHIASIS DUE TO IDIOPATHIC HYPERCALCIURIA IN A 12 YEAR OLD BOY: CASE REPORT

A. Christoforaki, **M.G. Avanidi**, A. Kleisarchaki,
V. Agelakou, G. Vlachaki, E. Kokkori

*Pediatrics, General Hospital of Venizelion,
Heraklion, Greece*

Introduction: Nephrolithiasis is recognized increasingly in children and inherited metabolic diseases are identified more frequently in children than in adults. The evaluation of a child with nephrolithiasis should be directed towards identifying physicochemical, anatomic and genetic factors predisposing to nephrolithiasis. Hypercalciuria is the most common metabolic cause of pediatric urinary calculi. In children with hypercalciuria the prevalence of urolithiasis in the family is 46-69%.

Methods: There is described the case of a 12-years-old non-Greek boy with short stature and history of recurrent nephrolithiasis since the age of 4 years old without further evaluation. Endocrinology tests for GH were normal and the bone age was markedly delayed. The kidneys ultrasound showed expanded nephrolithiasis. Hypercalciuria and hypocitraturia were revealed with normal serum calcium concentrations. In 24hours urine collection, oxalate was normal. PTH was normal. Bone mineral density was decreased.