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RENAL TUBULAR FUNCTION IN MINOR THALASSEMIA

S. Sadeghi-Bojd¹, M. Hashemi²

¹Pediatric, ²Pediatric Nephrology, Zhedan Medical Sciences of University, Zahedan, Iran

β - Thalassemia minor is a common heterozygous hemoglobinopathy that is characterized by both microcytosis and hypochromia. It has been postulated that low grade hemolysis, tubular iron deposition and toxins derived from erythrocytes might cause renal tubular damage in adult patients with β - thalassemia minor.

The aim of this study is to investigate the renal tubular function in children with β - thalassemia minor and to determine its possible harmful effects.

Methods:

The study was conducted on 50 children (22 male and 28 female) at the age of years (range 4-19 years) with β - Thalassemia minor. A control group was formed with 50 healthy children whose ages and sexes match those in the first group.

Blood and 24-hour urine samples were obtained for hematologic and biochemical analysis.

Results: There was statistically significant difference among the two groups in terms of the results of FE_{UA} (%), TPR (%), FE_k (%), GFR, Urine uric Acid, serum Mg ($P < 0/05$). But other significant signs of renal tubulopathy such as hypercalciuria, and tubular proteinuria (β_2 - microglobulinuria) were not seen.

Conclusion:

On the contrary of children with β - Thalassemia major, renal tubular dysfunction is not common in children with β - Thalassemia minor.

Also it suggested that in future studies to confirm renal tubular dysfunction should be considered specific test such as NAG measurement and specific parameter of oxidative stress such as urine zinc level (indirect evidence of oxidative stress).

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PROCALCITONIN AS EARLY MARKER IN UPPER URINARY TRACT INFECTION

S. Sadeghi-Bojd¹, M. Hashemi²

¹Pediatric Nephrology, ²Biochemistry, Zahedan Medical Sciences of University, Zahedan, Iran

In order to establish the most reliable marker for distinguishing urinary tract infection, we recorded the clinical features and admission leukocyte count, ESR, CRP and procalcitonin in 80 children aged 2 months-10 years admitted with a first episode of UTI. Fifty children with mean age 4/89 and 30 children with mean age 5/20 were compared. ESR, WBC and PCT but no CRP were significantly higher in patients with upper UTI ($p < 0/0001$). PCT had the best performance (0/5 ng) with sensitivity, specificity and positive and negative predictive value of %63, %83/3, %87/8 and 56/4/1 respectively. PCT cut-off value 2 ng/ml also had sensitivity, specificity and positive and negative value of %50, %96/6, %96/2 and %53/7 respectively. PCT was more sensitive and specific for the diagnosis of upper versus lower urinary tract infection than CRP. Serum PCT is a better marker than CRP for early prediction of pyelonephritis in children with a first episode of UTI.

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PULSE CYCLOPHOSPHAMIDE THERAPY FOR STEROID-RESISTANT FOCAL SEGMENTAL GLOMERULOSCLEROSIS IN CHILDREN

A. Sharipov, K. Khamzayev, B. Mamatkulov

Pediatric Nephrology, Tashkent Pediatric Medical Institute, Tashkent, Uzbekistan

Background: In children, steroid-resistant nephritic syndrome due to focal segmental glomerulosclerosis (FSGS) is frequently a progressive condition resulting in end-stage renal disease (ESRD). We report the response of 25 patients with steroid-resistant FSGS to treatment with intravenous pulse cyclophosphamide. Seven patients had initial steroid resistance and eighteen patients had late steroid resistance.

Materials and Methods: All patients were treated with intravenous pulse cyclophosphamide at a dose of 20 mg/kg/month for 12 months. Peroral prednisolone was given at a dose of 60 mg/m²/day for 6 weeks followed by 40 mg/m² on alternate days for 6 weeks and then tapered over next 6 weeks.

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