1648 ADENYLATE KINASE ACTIVITY IN THE KIDNEY OF DEVELOPING GUINEA PIG. Adarsh M. Kumar and Adrian Spitzer, Albert Einstein College of Medicine, Department of Pediatrics, Bronx, New York.

Experiments performed in our laboratories have revealed that the equilibrium constant, an expression of the relationship between the rate of synthesis and catabolism of ATP, is significantly higher in the newborn than in the adult. This could be due either to a higher turnover rate or to a rate limiting step in the generation of ATP. In order to test the latter alternative, we performed measurements of adenylate kinase, the enzyme which catalyses the phosphorylation of ADP to ATP. The measurements were done on kidneys obtained from guinea pigs (4-7 days and 6-8 weeks of age). Tissue extract was obtained by homogenizing the kidney and centrifuging it at 100,000 x g. The supernate was used for the enzyme assay. Activity was measured spectrophotometrically by coupling ADP with glucose, hexokinase, glucose-6-phosphate-dehydrogenase and NADP and monitoring formation of NADPH at 340 mμ. The results (mean ±SE) are expressed in μmoles ATP formed/min (unit) per mg protein.

	n	Units/mg protein
Newborn	5	1.91 ± 0.17
Adult	5	2.25 ± 0.23
P		>0.1

The results demonstrate that the amount of adenylate kinase per mg of protein is similar in the newborn and the adult guinea pig. Thus, the higher equilibrium constant observed in the kidney of the newborn should reflect a higher turnover rate of the adenine nucleotides.

1649 EARLY DIALYSIS AND FRESH FROZEN PLASMA (FFP) IN THE THERAPY OF HEMOLYTIC UREMIC SYNDROME (HUS). Roger E. Spitzer, State University of New York, Upstate Medical Center, Department of Pediatrics, Syracuse, NY.

In a 10 week period, 5 children (ages 21 months to 6 years) were seen with characteristic findings of HUS. All 5 presented with bloody diarrhea (3-8 days duration), azotemia (BUN:65-225 and creatinine:4.0-15.1), severe microangiopathic hemolytic anemia (Hgb:4.9-7.4 g%), and thrombocytopenia (platelet count: 13-64,000). Two patients had seizures. Multiple cultures for viruses and bacteria were all negative. Three children were anuric (14, 16 and 21 days) while 2 were severly oliguric (6 and 8 days). All five were treated with peritoneal dialysis from the time of admission and given packed cell transfusions as needed. Administration of FFP was started within the first 30 hours. The time for the platelet count to normalize appeared to vary with the volume of FFP given (0 cc/kg, 8 days; 30 cc/kg, 7 days; 40 cc/kg, 6 days; 66 cc/kg, 72 hours; 78 cc/kg, 48 hours). Neither the amount of FFP given nor the normal platelet count, however, appeared to influence the renal failure or the subsequent recovery of satisfactory renal function. None of the 5 children now require dialysis but 4 of 5 have reduced creatinine clearances (100, 60, 50, 50, 25 cc/minute). The degree of residual renal insufficiency relates well to the length of time these patients were anuric or oliguric (6, 8, 14, 16, and 21 days respectively). These data suggest that vigorous therapy with FFP may ameliorate platelet consumption in HUS and, coupled with early dialysis, be important for a satisfactory outcome.

†1650 C3NeF PRODUCTION BY PERIPHERAL BLOOD LYMPHOCYTES (PBL) FROM NORMAL INDIVIDUALS AND PATIENTS WITH MEMBRANO-PROLIFERATIVE GLOMERULONEPHRITIS (MPGN). Roger E. Spitzer and Ann E. Stitzel, S.U.N.Y., Upstate Medical Center, Dept. of Peds., Syracuse, NY.

C3NeF is an IgG antibody which stabilizes Alternative Pathway C3/C5 convertase activity (C3bBb) in patients with MPGN. To study the production of C3NeF, PBL from normals and 2 patients with MPGN were cultured in fetal calf serum with pokeweed mitogen for 14 days. The resultant supernatants, after adsorption, were added to sheep erythrocytes bearing C3bBb (EC3bBb). The cells were then quantitated for bound human IgG (C3NeF) by ELISA and decayed at 30° with measurement of residual convertase activity. Cells reacted with supernatants from MPGN patients contained 7-11 ng C3NeF (282-443 molecules IgG/cell) and after 10 minutes of decay had a residual convertase activity of 32-41% of the starting value (Control =14-18%). Cells reacted with supernatants from normals contained 4-9 ng C3NeF and had a residual convertase activity of 21-23%. Normal lymphocytes depleted of OK+T8 cells by monoclonal Ab adsorption yielded cultures which deposited 12-18 ng C3NeF and increased the residual convertase activity to 39%. Finally, IgG purified from normal serum was able to deposit 352 ng with a residual activity of 49%. Purified C3NeF deposited 1469 ng with residual convertase activity of 67%. These data indicate that C3NeF is produced in normal individuals under the control of OK +T8 cells. The increase in concentration of C3NeF in patients with MPGN may be the result of an alteration in this regulatory mechanism.

†1651 HYPERCALCIURIA: AN IMPORTANT CAUSE OF HEMATURIA IN CHILDREN. Fielding B. Stapleton, Shane Roy, III, Horace N. Noe, and Gerald Jerkins. Depts. of Peds. and Urol., Univ. TN. Ctr. Health Sci., and LeBonheur Children's Medical Center, Memphis, TN.

Hematuria may precede urolithiasis in children with hypercalciuria (HCU). To determine the incidence and nature of HCU in children with hematuria, urinary calcium excretion was examined in 83 consecutive patients with hematuria without urolithiasis, proteinuria, infection, sickle cell disease or systemic disease. HCU (urinary calcium >4mg/kg/d) was present in 23 children.

	HCU	Normal Calcium Excretion	P
n	23	60	
age, yrs	7.94±0.64	8.88±0.43	NS
Sex, f/m	8/15	24/36	NS
Gross hematuria	16(70%)	21(35%)	0.01
Family urolithiasis	17(77%)	9(15%)	0.001
Urine calcium, mg/kg	5.82±0.36	1.63±0.84	0.001

Oral calcium loading tests in HCU patients revealed absorptive HCU in 10 and renal HCU in 13. Serum PTH and bicarbonate concentrations were normal in all children with HCU. Hematuria resolved with anticalciuric therapy in 20 of 23 patients. Two children with HCU developed urolithiasis. In 4 patients with HCU, renal histology was normal in 3 and 1 patient had IgA nephropathy. No etiology of hematuria was found in 39 of 60 children with unexplained hematuria. We conclude that HCU occurs frequently in children with hematuria and that assessment of urinary calcium excretion is indicated in the evaluation of hematuria.

1652 LONG-TERM PROGNOSIS FOR CHILDREN WITH NEPHROTIC SYNDROME

Susan R. Wynn (Spon. by Gunnar B. Stickler), Mayo Graduate School of Medicine, Rochester, Minnesota.

One hundred fifty children with nephrotic syndrome were diagnosed between 1951 and 1967. Of 132 for whom information was available after 16 to 32 yr, 35 had died (mortality, 27%). Recurring edema and proteinuria had persisted in 15%. Three patients currently are on hemodialysis, and 22 patients died in renal failure which occurred 3 months to 8 years (mean, 3 years) after clinical onset. There were two deaths from complications of the nephrosis or the steroid therapy.

Hematuria occurred in 38% of the patients who died compared with 15% of those still alive. Hypertension also was more common at presentation among those who died (29% vs. 11%). Steroid resistance (no relief of edema or proteinuria after 6 weeks of therapy) was the most important feature predictive of poor outcome; all nine such patients died or are on dialysis.

The recurrence of edema, proteinuria, or steroid dependence into adulthood bore no relationship to the presenting symptoms of hematuria, hypertension, azotemia, or onset at less than 1 year of age or beyond 8 years. There was no increased incidence of malignancy, atopic disease, cardiovascular disease, or T-lymphocyte-related disorders. Of the 10 males who had received cyclophosphamide therapy, 4 have fathered children, but 1 who received chlorambucil has developed leukemia.

†1653 URINARY AND RENAL HISTOLOGICAL CHANGES IN CHILDREN WITH ACQUIRED IMMUNODEFICIENCY SYNDROME (AIDS). Jose Strauss, Brenda Montane, Gwendolyn Scott, Michael Freundlich, Carolyn Abitbol, Gaston Zilleruelo, Victoriano Pardo. Departments of Pediatrics and Pathology, University of Miami, Miami, Florida.

Patients with AIDS frequently develop circulating immune complexes (CIC) and elevated serum immunoglobulins which might predispose to renal disease. The latter has been recently described in >10% of adults with AIDS, but no such association has been reported in children. We studied 15 patients ($\bar{x}$ age 11 months, range 2-24) with proven AIDS (defined by an inverted T4/T8 ratio plus Kaposi's Sarcoma or unusual infections). All evidenced intermittent proteinuria (up to 55 mg/kg/day) and 7 had hematuria (up to 50 RBC's/HPF) and/or casts. Serum creatinine was normal ($\bar{x}$ 0.6 mg/dl). Elevated ESR (max. 116 mm/hr) was noted in all patients, increased CIC (C1Q and Raji) in 6 and high serum IgG (max. 4,910 mg/dl) in 10. Serum C3 and C4 were normal in all. Renal autopsy material was available in 7 patients of which 3 had abnormal findings: 1 with markedly increased mesangial matrix and nuclei, 1 with profuse electron-dense deposits in the mesangium, and 1 with immunofluorescent deposits of IgM and C3 in the mesangium. We conclude that AIDS is often associated with urinary and renal histological changes. These changes are not as severe as those in adults possibly due to the shorter duration of the disease, lack of associated complicating factors (drug addiction, etc.,) or to the renal and immune characteristics of children; still they may carry prognostic significance.