**1123** A USEFUL BIOCHEMICAL MARKER IN THE IDENTIFICATION OF HEREDITARY NEPHRITIS: HYDROXYLYSINE GLYCOSIDES (OHLG's). L.U. Tina, M. Lou, D. DiZio and P.L. Calcagno. Department of Pediatrics, Georgetown University School of Medicine, Washington, D.C. and Alfred I. DuPont Institute, Department of Biochemistry, Wilmington, Delaware.

Collagen metabolism was studied in 9 children with hereditary nephritis and 10 family members with microscopic hematuria. The diagnosis was confirmed in all 9 children by percutaneous renal biopsy. No previous studies exist defining the collagen turnover via the utilization of the urinary OHLG's, hydroxylysyl-galactosyl-glucose (HGG) and hydroxylysyl-galactose (HG), in hereditary nephritis. Mean urinary OHLG's values in the biopsy group was 113 u M/g creatinine as compared to 15 control children of 74 u M/g creatinine. The mean urinary values of 128 u M/g creatinine for the siblings were comparable to the nephritis group.

Two additional patients with the Nail-Patella Syndrome also demonstrated increased urinary excretion of OHLG's. The significantly increased urinary excretion of these metabolites, HGG and HG, support the thesis of an altered collagen disorder.

This study suggests that urinary hydroxylysine glycosides may be used as a useful marker in the recognition of the early stage of hereditary nephritis.

**1124** NON-RENAL INVOLVEMENT IN HEMOLYTIC UREMIC SYNDROME. Kirti Upadhyaya, Kenneth Barwick, Mark Fishaut and Norman J. Siegel, Depts. of Ped. & Path., Yale Sch. of Med., New Haven, Conn.

Over the past several years hemolytic uremic syndrome has been considered an intravascular coagulopathy which is localized to the kidneys. The clinical outcome and histopathologic findings in 12 children who presented with microangiopathic hemolytic anemia, thrombocytopenia, and azotemia are the basis of this report. Renal failure was recognized early and appropriate therapy instituted in all cases. Seven patients recovered completely after a short period of oliguria. Two patients had severe renal failure and required bilateral nephrectomy for control of HBP: both have been transplanted and have normal renal function one and three years later. Three patients died within 8 days of diagnosis: all 3 had been anuric with unresponsive seizures despite early dialysis. Histologic examination in these 3 pts revealed fibrin thrombi and/or infarcts in brain, colon, heart, liver, thyroid, lymph node, and adrenal glands. Ischemic colitis and brain infarcts were notably prevalent.

With early and appropriate therapy for renal failure, the mortality in hemolytic uremic syndrome may be determined by the degree of non-renal involvement, and supportive care may unmask previously unappreciated systemic manifestations of hemolytic uremic syndrome.

**1125** FAMILIAL NEPHRITIS, A VARIANT OF ALPORT'S SYNDROME? Nina B. Vicente, Dinyar B. Bhathena and Nancy H. Holland. Depts. of Ped. and Path. University of Ky. Medical Center, Lexington, Kentucky.

A form of familial nephritis with many features distinct from Alport's Syndrome (AS) was diagnosed in 7 of 10 children of clinically unaffected parents. The disease is manifested by progressive renal failure without antecedent hematuria, proteinuria or discernible auditory and ocular defects. The disease is more severe in females, 3 of 4 affected reaching end-stage by age 12 years. Of 3 affected males, only one 17 year old has developed chronic renal failure.

Biopsies in 3 of 4 clinically unaffected siblings, 3 female and 1 male, showed focal glomerular as well as interstitial changes with normal vasculature; no immune complex deposits were seen. The chief ultrastructural features were 1) thinning of several peripheral glomerular capillary basement membranes (GBM) in all 2) irregular GBM thickening with small lucent areas in a few loops 3) small podocytes frequently sprouting foot processes directly from scant perniculear cytoplasm. The characteristic changes of AS were not seen in 3. However, in the fourth patient one isolated loop showed changes that resembled or were compatible with those of AS.

Because of the absence of a family history, the asymptomatic progressive renal failure particularly in young females and some unusual ultrastructural features, we suggest this family represents either a variant of AS or an undescribed form of familial glomerulonephritis.

**1126** MYCOPLASMA PNEUMONIA,PLEURAL EFFUSION AND ACUTE GLOMERULONEPHRITIS.Benedetto B.Vitullo,Sean O'Regan, Jean-Pierre de Chadarevian and Bernard S.Kaplan. McGill Univ.-Montreal Children's Hosp.Research Inst.,Depts.of Nephrology,and Pathology,Montreal,Canada.

An 11-year old girl presented with pneumonia,pleural effusion,hematuria and red blood cell casts.The blood pressure,serum electrolytes,immunoglobulins and C3 concentrations were within normal range.Sputum cultures were negative for M.pneumoniae.The ASO titers were less than 200 Todd U. Cold agglutinin titers were 1/64 on admission and 1/256 on Day 9.Complement fixation titers for M.pneumoniae were negative on admission;subsequent titers measured on several occasions were 1/128.A thoracentesis revealed straw-colored fluid;gram stain and cultures of the pleural fluid for bacteria,viruses,fungi and acid-fast bacilli were negative.A percutaneous renal biopsy revealed an acute glomerulonephritis by light microscopy;immunofluorescent studies showed deposits of IgG,C3 and mycoplasma antigen along the glomerular capillary walls and in the mesangium;subendothelial and subepithelial deposits were seen by electron microscopy.Circulating immune complexes,19S or larger,have been demonstrated in patients with acute respiratory illness caused by M.pneumoniae when clinical signs were most pronounced.

This is the first report of immune complexes(antigen-antibody and C3)deposited in the kidney of a patient with M.pneumoniae infection and glomerulonephritis.

**1127** SCREENING FOR HYPERTENSION IN UNIVERSITY OF FLORIDA STUDENTS. Robert L. Williams, Carl D. Sorgen, Eduardo H. Garin, George A. Richard (Spon. by Gerold L. Schiebler) Univ. of Florida, College of Medicine, Department of Pediatrics, Gainesville.

College Students aged 18-30 years were screened for hypertension at the student infirmary. 550 students had a single blood pressure reading taken in the sitting position. 50 students (9%) fulfilled our definition of high blood pressure by having either a systolic pressure equal to or greater than 140 mm/Hg or a diastolic pressure equal to or greater than 90 mm/Hg. Of these students with abnormal blood pressures, 34 returned for 2 further determinations. 22/34 (65%) demonstrated persistent hypertension during each of the three visits. Of those with persistent hypertension, 6/22 (27%) had an elevation of the systolic pressure, 4/22 (18%) had an elevation of the diastolic pressure and 12/22 (54%) had an elevation of both when initially screened. Of the 12 students without persistence of hypertension (labile), 3/12 (25%) had an elevated systolic pressure, 4/22 (33%) had an elevated diastolic pressure, and 5/22 (43%) had an elevation of both.

Only 3/22 (14%) of the students with persistent and none with labile hypertension had a diastolic pressure equal to or greater than 105 mm/Hg on the initial screening pressure, indicating hypertension is usually mild in this age group. The type of hypertension (systolic, diastolic, or both) does not allow the identification of those individuals who have labile or persistent hypertension.

**1128** SEIZURES: A SIGNIFICANT SIDE EFFECT OF CHLORAMBUCIL THERAPY. Williams, Susan A., Makker, Sudesh P., and Grupe, Warren E., Depts of Peds., Children's Hospital Medical Center, Boston, and Case Western Reserve University, Cleveland.

Of 88 children with nephrotic syndrome treated with 96 courses of chlorambucil and prednisone, 6 (3 males, 3 females) experienced seizures during therapy. Diagnoses by renal biopsy were: focal segmental glomerulosclerosis (3), mesangial proliferative variant (1) and minimal lesion (1); one steroid responsive patient was not biopsied. The age at onset of disease ranged from 1.1-6.2 yrs, duration of illness from 0.4-8.5 yrs. The dose of chlorambucil varied between 0.18-0.67mg/kg/d for 8 days-13 weeks of treatment (total dose 1.6-36.7mg/kg).

In all, seizures were initially focal with progression to generalized myoclonus; 3 had multiple seizures while on therapy. No child had previous seizures while on steroid therapy alone except one who had a seizure with hypertensive encephalopathy 4 yrs previously. No child had hypertension, fever or metabolic abnormality at the time of seizures. Lumbar puncture in 4 was within normal limits. EEG showed focal changes in 5 and diffuse slowing in one; follow-up EEG in 4 had returned to normal. No child had persistence of seizure activity after chlorambucil discontinued.

Even though seizure activity can not be related to age, duration of illness, sex, dosage, duration of treatment, or underlying disease, chlorambucil appears to be implicated as a contributing factor to these seizures.