1075 DIFFUSE ARTERIOVENOUS (AV) MALFORMATION OF BOTH KIDNEYS Bernard G. Gauthier, Edward S. Wind, Ashok C. Shende, Philip Lanzkowsky, George P. Pillari, Sch. of Med. Health Sciences Ctr. State University of N.Y. at Stony Brook and Long Island Jewish-Hillside Medical Ctr. Depts. of Pediatrics and Radiology, New Hyde Park, N.Y.

AV malformations of the kidneys are rare and AV malformations replacing the normal vasculature of both kidneys, so far as we are aware, has not been previously described. The purpose of this abstract is to describe such an abnormality in a girl aged 3 years and 10 months. She presented with hypertension (190/140mmHg), a hemolytic anemia with red cell fragmentation (Hb 6.1 G/dl, Reticulocytes 6.2%,haptoglobin 0 mg/dl), thrombocytopenia (platelets 72000/cmm) and decreased renal function (S.Creatinine 1.4 mg/dl, BUN 32 mg/dl, GFR 35ml/min/1.73m^2). There was no evidence of intravascular coagulation. Blood renin was normal. She also had hypertrophy of the left leg, with prominent veins, probably due to an associated AV malformation of the leg. Arteriography showed that the vasculature throughout both kidneys, was replaced by multiple AV malformations. Drainage was through large, tortuous veins anastomosing with periureteral and pelvic veins. The inferior vena cava was visualized early. She has been treated with chlorothiazide, propranolol, hydrallazine, methyldopa, aspirin and dipyridamole for 9 months. The hypertension is controlled, and the platelet count has returned to normal. The Hb has risen to 10.5 G/dl but Red cell fragmentation persists. The GFR has risen to only 57 ml/min/1.73m^2.

1076 MESANGIAL PROLIFERATIVE GLOMERULONEPHRITIS PRESENTING AS NEPHROTIC SYNDROME IN CHILDHOOD. Joseph Gangiacomo and Cheng-Chang Tsai. (Intr. by A. E. McElfresh) Cardinal Glennon Mem. Hosp. for Children and St. Louis Univ. Sch. of Med., Dept. of Ped. and Path., St. Louis, Missouri.

Despite two controlled investigations of childhood nephrosis totalling 373 patients, there remains a paucity of detailed clinical and pathologic information of mesangial proliferative glomerulonephritis with the nephrotic syndrome in childhood. We had the opportunity to study 8 patients with the nephrotic syndrome, the mean age of onset was 7.2 years with a range of 3.5 to 12.2 years; there were 5 males and 3 females. All patients had renal biopsies which were studied by light, electron and immunofluorescent microscopy. The pathologic findings with hematoxylin-eosin showed predominant mesangial proliferation in all patients. Immunofluorescences revealed positive mesangial fluorescence to immunoglobulin (Ig) M in 8 patients, 6 of 8 had C3 complement and fibrin/fibrinogen, 4 of 8 had IgG. Electron microscopy showed electron dense deposits in the mesangium.

Seven of the 8 patients were steroid sensitive and frequent relapsing nephrotics. One is steroid resistant for 1.5 years but renal function remained normal. It has been assumed that children with steroid sensitive nephrotic syndrome with the onset before 5 years of age have idiopathic nephrotic syndrome (I.N.S.), 2 of the present patients were less than 5 years at onset; therefore, this may no longer be an accurate assumption. Therefore previous studies of steroid sensitive nephrotic syndrome with onset less than 5 years without renal biopsies may not fulfill the pathologic criteria of I.N.S.

1077 PATHOGENETIC VARIABILITY OF NEPHROPATHY (N) IN JUVENILE DIABETES MELLITUS (JDM). Nisan Gilboa, Dennis P. Durante, Stephen J. Guggenheim, and Rawle M. McIntosh, University of Colorado Medical Center, Dept. of Peds., Denver, Colorado.

Clinicopathologic studies of 4 patients with JDM and N demonstrated the pathogenetic variability of the nephropathy. Only 1 patient had the typical diabetic N in form of proteinuria (P), hypertension (H), decreased glomerular filtration rate (GFR) and diffuse and nodular diabetic glomerulosclerosis (DG). In another patient the N had the features of steroid responsive nephrotic syndrome (SRNS) of childhood superimposed on a very mild DG. This patient had HLA-B$_{12}$ antigen associated with SRNS. The third patient also had a nephrotic syndrome (NS) which, unlike the other, responded only partially to treatment. In addition, she had Grave's disease (GD). Kidney biopsy 9 years after appearance of the NS showed only mild DG. The presence of HLA-B$_8$ antigen, shown to be associated with SRNS, JDM and GD could be suggestive of a common pathogenesis of all 3 disorders in this patient. In the fourth patient, the N in form of H, P and rapidly declining GFR could have been related more to the severe tubulointerstitial disease of unknown etiology than to the moderate DG. The presence of renal tubular epithelial antigen in the mesangium in this case, may have had some pathogenetic significance. The poor prognosis of diabetic N warrants a careful search for other potentially treatable, causes of N in patients with JDM.

1078 ELEVATED PLASMA RENIN ACTIVITY AND "BIG RENIN" IN MESOBLASTIC NEPHROMA. Ted D. Groshong, Judith H. Miles, John H. Bauer, Myron Weinberger, and Nasrollah Hakami. (Spon. by Calvin Woodruff). Departments of Child Health and Medicine of the University of Missouri, Columbia, Missouri and Department of Medicine, Indiana University, Indianapolis, Indiana.

Elevation of plasma renin activity (PRA) has been reported with several kidney tumors, but not with mesoblastic nephroma (MN), a common renal tumor of early infancy. We have studied a 3 month old infant with a large intrarenal mass, histologically characterized as MN. Prior to removal of the tumor, the infant exhibited severe hypertension, and moderate hypokalemia. PRA from peripheral veins, aorta, and inferior vena cava, below the renal vein were markedly elevated to greater than 300 ng/ml/hour. Massive amounts of "Big Renin", greater than 1000 ng/ml/3 hour were also demonstrated. Normalization of blood pressure and serum potassium occurred shortly after removal of the renal mass, and PRA and "Big Renin" returned to basal levels. There is no evidence of tumor recurrence and serial measurements of PRA over a 12 month period shows levels below 5 ng/ml/hour and "Big Renin" remains undetectable. Also, in repeated studies, expansion of plasma volume resulted in suppression of PRA, demonstrating normal regulation of renin release. These observations suggest: (1) Mesoblastic nephroma may be associated with autonomous production of PRA, producing hypertension and hypokalemia; and (2) Serial measurements of PRA may be a useful biologic marker for detection of early recurrence of the tumor.

1079 VALUE OF ASO TITERS FOR DIFFERENTIAL DIAGNOSIS OF RENAL DISEASES IN CHILDREN. Walter Heymann, M.D. and Bernard Boxerbaum, M.D. Department of Pediatrics, Case Western Reserve University and University Hospitals, Cleveland, Ohio.

A review of ASO titers in 87 children with various renal diseases makes it probable that ASO titers are of significance in the differential diagnosis of pediatric renal diseases. Values obtained in cases of acute post-infectious glomerulonephritis were, as is known, elevated, even in cases which were associated with a nephrotic syndrome. In contradistinction, every one of 35 cases of primary, idiopathic, nephrotic syndrome of childhood had values lower than 100. This was not noted in 16 cases of primary, chronic glomerulonephritis, even though 10 of them were observed during nephrotic phases of the disease. The low values in the nephrotic syndrome may be due to the hyperlipidemia or to the loss of ASO proteins in the proteinuric urine. This cannot be decided as yet and will require additional data.

Thus we believe that it is important to include a determination of ASO titers in the workup of every child with renal disease, even when we know that it has not been elicited by a preceding streptococcal infection.

The establishment of the diagnosis of "primary," "idiopathic" nephrotic syndrome of childhood demands additional studies if the ASO titer is above 100. The low titers have been observed within the first few weeks after onset and have been noted to last for many years of stable, complete remissions.

1080 DEFECTIVE CYCLIC AMP GENERATION FOLLOWING PITRESSIN HORMONE (ADH) INFUSION IN GIRLS WITH NEPHROGENIC DIABETES INSIPIDUS (NDI). Ronald J. Hogg and John W. Balfe, Univ. of Texas Southwestern Medical School, Dallas, Tx and Hospital for Sick Children, Toronto, Can. (Spon. by H.G. Worthen).

ADH exerts its effect on urinary volume and concentration via intra-cellular cyclic AMP generation. The integrity of this system can be studied by measuring urinary cAMP excretion following ADH administration. This was carried out by infusing ADH (0.5 to 1.0 mU/min) to 4 NDI girls in a state of water diuresis. These NDI girls presented during a 10 year period to a large children's hospital. For comparison, the same procedure was carried out in 2 normal adults, who showed 100% increase in UcAMP, and in 3 NDI boys who showed no increase in UcAMP.

	Urine volume ml/min/KgBWt		Urine Osm. mOsm/KgH$_2$O		UcAMP nMol/m^2/min.		UcAMP nMol/mg.creatinine	
	C	E	C	E	C	E	C	E
NDI-girls (n=4)	0.16	0.14	68	66	2.37	2.34	10.7	10.9
NDI-boys (n=3)	0.24	0.22	60	53	3.10	2.10	6.73	5.70
Controls (n=2)	0.07	0.01	198	826	1.01	1.93	1.46	3.32

C = control period. E = experimental period (ADH infusion)

The absence of any effect of ADH on UcAMP in girls differs from the result obtained in one previous case reported and demonstrates that females with this disorder are also afflicted with a deficient ADH-sensitive adenyl cyclase system.