EFFECT OF (Sar^1, Ile^8)-ANGIOTENSIN II INHIBITOR ON GLOMERULAR FILTRATION AND RENAL PLASMA FLOW IN THE FETAL LAMB. E.S. Moore, J.B. Paton, C.W. deLannoy, Y. Roan, E. Cevallos, M. Ocampo and E.C. Lyons. Michael Reese Medical Center, Dept. of Ped., Chicago, Illinois.

Fetal plasma renin activity (PRA) is high when compared to the ewe. To study the effect of this high PRA on fetal renal function, (Sar^1, Ile^8)-Angiotensin II (A-II) inhibitor was infused into 5 fetal lambs. After measuring GFR, C_{PAH}, filtration fraction (F.F.), urine flow (UV), Cosm, arterial pressure and renal vascular resistance for 3 control periods, A-II inhibitor was infused at 7 mgm/kg/min for 3 study periods. Two (2) recovery periods were then done.

	Control	A-II Inhibitor Infusion	Recovery
GFR+	4.56	2.64	2.58
C_{PAH}+	6.16	3.43	3.12
F.F.	0.78	0.80	0.83
UV	0.85	0.47	0.35
P.P.*	7	15	9

+Ml/min; *P.P. Pulse pressure

Systolic pressure did not fall but there was a significant increase in P.P. Inhibition of A-II resulted in a significant fall in GFR, C_{PAH} and UV while the F.F. increased. This effect persisted after discontinuing the infusion. These data suggest that elevated fetal PRA is an appropriate response to low total renal blood flow and that GFR, renal plasma flow and UV are significantly maintained by the high PRA.

RENAL FUNCTION IN LOW BIRTH WEIGHT (LBW) INFANTS. E. S. Moore, J. B. Paton, K. Tanprasert, E. Cevallos, Y. Roan and D. E. Fisher. Michael Reese Medical Center, Dept. of Ped., Chicago, Illinois. (Intr. by Samuel S. Spector).

Clearance studies were done in 66 healthy LBW infants in 6 weight groups: <1200 g (n-4); 1200-1700 g (n-22); 1701-2200 g (n-20); 2201-2700 g (n-10); 2701-3200 g (n-8) and >3200 g (n-2). Studies were done at age 1-2 days, 3-5 days and greater than 10 days. The following studies were done: UV, $C_{creatinine}$, C_{Na^+}, % T_{Na}, Cosm/GFR, C_{H_2O}/GFR, $U_{K}V$, $U_{Cl}V$, $U_{TA}V$, $U_{NH_4}V$, U_{osm}, Sp. Gr., U_{pH} and $U_{proteinV}$. Urine flow (ml/hr/kg) during the first 10 days of life was directly related to body weight rather than to volume of fluid intake. Endogenous C_{creat} tended to be higher in the first 48 hrs than at 5 and 10 days even though more fluids were taken during the latter periods. Uosms were low and varied directly with weight initially, and decreasing as more fluids were given. Infants <1200 gms never produced urines with a Sp. Gr. >1.005 (Uosm-160). Fractional T_{Na} was the same in all groups (99.85%). However, Cosm/GFR and C_{H_2O}/GFR varied directly with weight and increased fluid intake. Spontaneous acidification of the urine was by excretion of TA alone as NH_4 was not produced by any group. These studies suggest that qualitative renal tubular function in LBW infants is similar for all weight groups during the first month of life. Quantitative differences of urine flow, GFR, concentrating capacity etc. are related to body weight and to volume of fluid intake rather than to differences in degree of maturation of function.

SERUM TOTAL HEMOLYTIC COMPLEMENT (CH50) LEVELS IN NORMAL INFANTS AND CHILDREN, Michael E. Norman, Arlene Taylor, Ulf R. Nilsson, Eric P. Gall and Larry Laster, Univ. of Penn., Depts. Ped., Med. and Dent., Philadelphia

The purpose of this study was to establish normal CH50 levels in healthy infants and children, and to compare them with CH50 levels in a population of hospitalized children (HC), and children with acute, poststreptococcal, membrano-proliferative and Lupus glomerulonephritis (GN). A modification of Kabat and Mayer's method was used; serum was examined fresh or within 2 weeks of quick freezing and storage at -70°C.

AGE GROUP	NUMBER	MALE/FEMALE	BL/WH	CH50(u/ml)
cord blood	24	12 12	13 11	120±63(2st.dev.)
2 - 7 days	23	11 12	12 11	152±53
1 - 5 months	17	8 9	4 13	146±76
6 - 23 months	25	13 12	12 13	151±53
2 - 5 years	27	14 13	12 15	134±57
6 - 9 years	28	17 11	15 13	150±64
10 - 14 years	23	17 6	10 13	143±56
TOTAL (normal):	167	92 75	78 89	142±64
HC 1 mos-17 years:	51	33 18	27 24	156±88
Children with GN:	13	7 6	7 6	48±118

In normal children, no correlation was found between CH50 levels and age, sex or race. Mean CH50 titers were similar in normal and hospitalized children, but statistically significantly different from the mean titer in children with GN ($p < .001$). In screening children with illnesses other than GN, the CH50 test has limited diagnostic value.

PROGNOSIS OF GLOMERULAR FOCAL SCLEROSIS IN CHILDREN. Hermann Olbing, Martin A. Nash, Jay Bernstein, Boyce Bennett, Adrian Spitzer, and Ira Greifer (Intr. by Chester M. Edelmann, Jr.), Dept. of Ped. and Path., A. Einstein Col. of Med., Bronx, N.Y.

It is generally stated that focal sclerotic lesions of the glomeruli are associated with a poor prognosis. No distinction is usually made between focal global sclerosis (complete hyalinization of affected glomeruli) and focal segmental sclerosis (hyalinization of only one area of the affected glomeruli). Seventeen children with focal sclerosis were surveyed to determine whether the type of lesion has prognostic significance. Four children, 3 male 1 female, presenting with nephrotic syndrome (NS) at the age of 13 to 19 months, were found to have focal global sclerosis. They all responded to prednisone therapy, relapsed frequently, and have normal renal function 9 to 14 years later. The 13 children with focal segmental sclerosis ranged in age from 1½ to 13 years; 7 were male and 6 female. Eleven presented with NS. Of these, 2 have died, 2 have received renal transplants, and 5 have mild to moderate decreases in renal function and show progression on subsequent biopsies. It is suggested that: 1) Focal global sclerosis is a disorder different from focal segmental sclerosis, presenting at an earlier age, responding to steroid therapy, and carrying a good prognosis. 2) Although focal segmental sclerosis is usually associated with NS, it can also present with only proteinuria and hematuria. Patients with this lesion do not respond to steroid therapy. Despite the fact that some patients maintain near-normal renal function over several years, all show histological deterioration.

LATE-ONSET NEPHROPATHIC CYSTINOSIS WITH MASSIVE PROTEINURIA. A STUDY OF THE RENAL LESIONS AND TUBULAR FUNCTIONS. Rufino C. Pabico, Michael F. Bryson, Barbara McKenna, Bernard Panner, Richard B. Freeman, (Intr. by Gilbert B. Forbes). Univ. of Rochester Med.Ctr., Depts. of Med., Ped., and Path., Rochester, N.Y.

Cystinosis has not been associated with massive proteinuria. Although tubular dysfunctions are frequently noted, data on tubular kinetics prior to the onset of renal failure are lacking. Glomerular filtration rate(C_{in}), renal plasma flow(C_{PAH}), urinary concentration(U/Posm), amino acid excretion, maximum tubular reabsorption of glucose(T_{mG}) and secretion of PAH (T_{mPAH}), and urinary acidification were measured in 3 siblings (2 boys 12 and 18 yrs and a 9 yr old girl) with cystinosis and massive proteinuria(6.0, 13.0 and 2.0 Gm/day). Renal biopsy was performed in all. Light and electron-microscopy and immunofluorescent studies were done. The results are tabulated:

Patients	1	2	3
C_{in} (ml/min)	104	41	141
C_{PAH} (ml/min)	396	119	536
F.F.	0.25	0.33	0.26
U/P osm	2.3	2.2	3.3
Tm_G (mg/min)	139	45	355
Tm_G/Cin	1.37	1.10	2.52
Tm_{PAH} (mg/min)	32	15	45
Tm_{PAH}/Cin	0.31	0.37	0.33

Despite normal C_{in} in patients 1 and 3, tubular functions were already impaired. Patient 2 had the worse function. Phosphaturia, aminoaciduria and failure to acidify were noted. The biopsy showed glomerular sclerosis in all patients and exudative lesions in patient 2. No immune complex deposits were seen. Cystine crystals were present in tubular cells, in the lumen and interstitium.

HYPERTENSION IN INFANTS. Lauren B. Plumer, George W. Kaplan and Stanley A. Mendoza. Univ. of Calif., San Diego, Depts of Ped and Surgery/Urology, La Jolla.

Ten infants less than three months of age were found to have severe hypertension (140 to 170 mm Hg systolic). Of the ten infants, four weighed less than 5 lbs. at birth, six had respiratory distress syndrome, three had patent ductus arteriosus, eight had hematuria, and eight had umbilical arterial catheters in place for 5 to 74 days.

Aortography demonstrated a cause of the hypertension in all 6 infants studied. Five of these studies demonstrated thrombi in the aorta and/or the renal arteries. The sixth infant had renal artery stenosis. Two additional infants had aortic and renal arterial thrombi demonstrated at autopsy. A variety of antihypertensive drugs were administered in high doses with a generally poor response. Five infants died. Three recovered spontaneously, including one with a documented renal artery thrombus and the two who were not studied. Two infants recovered following nephrectomy.

Hypertension in the young infant carries a high mortality rate. Response to medical management may be unsatisfactory. Aortography has a high yield and should be performed early followed by surgery if indicated.