1516 TREATMENT OF FOCAL GLOMERULOSCLEROSIS (FGS) WITH ALKYLATING AGENTS. Jacques Lemire, Jean-Pierre de Chadarévian and Bernard S. Kaplan. McGill University, The Montreal Children's Hospital, Departments of Nephrology and Pathology, Montreal, Canada.

Nephrotic syndrome secondary to FGS is usually unresponsive to steroids and progresses, in many cases, to end-stage renal disease. Alkylating agents, chlorambucil (CH), cyclophosphamide (CY), appear to decrease the frequency of relapses or to prolong remission in the minimal change nephrotic syndrome (MCNS), but their efficacy in the treatment of FGS is still debatable.

5 patients presented with nephrotic syndrome. Initially responsive to prednisone, 4 of 5 had multiple relapses, but at the time of treatment with alkylating agents, steroid resistance was present in each (1 to 3 months duration).

Patient	P.G.	J.P.B.	P.P.	G.F.	B.B.
Age of Onset of NS (Yr.)/Sex	1.8/F	1.4/M	3.9/F	3.7/F	7/F
Treatment: Age (Yr.)	4.4	2.3	7.1	4.1	7.2
Agent	CH	CH	CH	CY	CY
Duration (Mo.)	3	2	2	2.5	2.5

Percutaneous renal biopsy was done before therapy; each had FGS. Interstitial fibrosis and tubular atrophy were present in G.F. CH, 0.3 mg/k/day or CY, 1.5 mg/kg/day were started and prednisone 2 mg/kg/day was continued every second day. A complete remission was obtained in every patient and maintained as long as 21 m after cessation of the alkylating therapy.

Some patients with FGS thus appear to benefit from 8 w treatment with an alkylating agent.

1517 PROLONGED USE OF INDOMETHACIN IN CYSTINOSIS. Jacques Lemire and Bernard S. Kaplan. McGill University, The Montreal Children's Hospital, Department of Nephrology, Montreal, Canada.

Indomethacin improves the polyuria associated with Fanconi syndrome in patients with cystinosis. However, its use may be vitiated by its deleterious effect on GFR.

J.B. presented at 4 y with cystinosis. After correction of the acid base and serum electrolyte abnormalities with supplemental therapy, indomethacin, 2 mg/kg/d was started. Urine output decreased from 1.9 to 0.9 l/d and she gained weight. 3 y later, serum creatinine suddenly increased from 2.3 to 3.8 mg/dl. After stopping indomethacin, the creatinine decreased to 2.5 mg/dl but polyuria reappeared.

H.T. was diagnosed at 14 m. Indomethacin, 2 mg/kg/d was instituted; urine output decreased from 2.3 to 1.3 l/d and she gained weight. 2 y later, serum creatinine rose from 1.4 to 2.6 mg/dl and the drug was stopped. 2 w later, creatinine was 1.7 mg/dl. Because of rapid reappearance of polyuria associated with irritability, indomethacin was re-started at 1 mg/kg/d. Urine output decreased and 2 m later, creatinine stabilized at 1.4 mg/dl.

Indomethacin is definitely effective in reducing polyuria in cystinotic patients, but can also produce a reversible increase in the serum creatinine concentration.

● **1518** IMPROVED TECHNIQUE FOR ASSESSING URINARY ACIDIFICATION Linda C. Loney, Laura L. Norling and Alan M. Robson. Dept. of Pediatrics, Washington Univ. School of Med. and St. Louis Children's Hospital, St. Louis, MO.

Ammonium chloride (NH_4Cl) loading to evaluate urinary acidification often is poorly tolerated because of severe side effects. Indeed it may produce severe uncompensated metabolic acidosis in infants (J Peds. 1977, 91:263). We have developed an alternate protocol utilizing the intravenous infusion of arginine hydrochloride (HCl) and have studied this in 22 children, of whom 4 had distal renal tubular acidosis (dRTA), 2 had proximal renal tubular acidosis (pRTA), 14 had recovered from glomerulonephritis and 2 had no evidence of renal disease. Arginine HCl was infused over 2.0 to 2.5 hours in a dose equivalent to between 100 to 150 mEq $[H^+]$ per m^2 SA in patients with RTA or decreased GFR and between 150 to 200 mEq $[H^+]$ per m^2 in the remainder. This resulted in a systemic acidosis sufficient to separate patients with dRTA from the remainder. In no one with dRTA was urine acidified below pH 5.50, lowest values being seen at 5 to 6 hr after starting the infusion. Urine pH was reduced to between 4.65 and 5.35 in all remaining subjects, values below 5.50 occurring in each within 3 hr of starting the infusion. In no instance did the systemic acidosis become excessive or induce symptoms. The procedure was well tolerated by the patients: the severe nausea, vomiting and cramping abdominal pain frequently associated with NH_4Cl loading did not occur. We recommend that arginine HCl, rather than NH_4Cl be used to study urinary acidification because of the safety, reliability, and better patient acceptance of the test.

1519 ROLE OF THE ANION IN VOLUME EXPANSION NATRIURESIS IN NEWBORN DOGS. J.M. Lorenz, L.I. Kleinman, T.A. Disney U. Cincinnati Coll. Med., Dept. Pediatrics, Cincinnati

Neonatal dogs 2 to 18 days had volume expansion (VE) with either NaCl (n=7) or $NaHCO_3$ (n=8) to determine the role of the anion in VE natriuresis. In VE with NaCl, Fractional Excretion (FE) of sodium, FENa, = 1.1%, FEK = 41.8%, FECl = 3.0%, $FEHCO_3$ = .03%. In VE with $NaHCO_3$, plasma HCO_3 was elevated (37.3 mM) and FENa = 2.8% ($p<.05$), FEK = 84.7% ($p<.01$), FECl = 0.6% ($p<.01$), $FEHCO_3$ = 7.3% ($p<.01$). When plasma HCO_3 was elevated (32.7 mM) without VE by exchange transfusion with high HCO_3, low Cl blood in 15 puppies, FENa = .4%, $FEHCO_3$ = .7%, FEK = 47.1%. Without VE in 14 puppies with normal plasma HCO_3, receiving ethacrynic acid and amiloride to block Henle's Loop (HL) and distal tubular function, FENa = 27%, $FEHCO_3$ = 2%, FEK = 30%, FECl = 37%, but with saline VE and these diuretics FENa = 55% ($p<.01$), FECl = 64.2% ($p<.01$), $FEHCO_3$ = 2.4% (N.S.), FEK = 68% ($p<.01$). We conclude that without VE almost all HCO_3 is reabsorbed in the proximal tubule (PT) even with high plasma HCO_3. VE, due either to NaCl or $NaHCO_3$, inhibits PT Na reabsorption, resulting in increased loads of Na, HCO_3 and Cl to HL. However, with VE due to NaCl relatively more Cl and less HCO_3 is delivered to HL than during VE with $NaHCO_3$. Thus, in the saline group HL reabsorbs more Na (as Cl) than in the HCO_3 group so that in the latter group, more Na is presented to the distal tubule (as HCO_3), resulting in greater distal K secretion. The net effect is greater Na, K and HCO_3 and less Cl excretion during VE with $NaHCO_3$ than with NaCl.

1520 PERCUTANEOUS TRANSLUMINAL ANGIOPLASTY IN TRANSPLANT RENAL ARTERY STENOSIS (RAS). Robert C. MacDonell, Jr, Amil J. Gerlock, Jr, H.Keith Johnson, Marion B. Tallent, Robert E. Richie, Bruce I. Turner, and Gary D. Niblack (Spon by Robert C. Boerth). Vanderbilt Univ Medical Center; Depts of Pediatrics, Medicine, Radiology, Surgery, Urology and Microbiology; Nashville, Tenn.

Hypertension(HT) due to RAS complicates 5-10% of renal transplants and jeopardizes graft function. Alternatives to surgery in immune suppressed transplant recipients are desirable. 4 subjects developed HT with hyperreninemia within 1 yr post transplant. Significant RAS was demonstrated angiographically in each. Therapy with percutaneous transluminal angioplasty produced:

CASE	AGE (yr)	RAS %	BLOOD PRESSURE (mmHg)		RENIN (ng/ml/hr)		CREATININE (mg/dl)		F/U (mo)	RX
			pre	*post*	*pre*	*post*	*pre*	*post*		
1	24	90	170/110	140/90	16	2	2.1	2.7	11	NONE
2	11	65	180/120	120/80	7	5	1.0	1.1	9	NONE
3	14	85	180/120	130/80	30	36	1.7	1.5	8	INCR
4	15	75	150/110	130/86	16	2	1.7	1/5	3	NONE

3 subjects remain normotensive, on no RX, 3 to 11 mos post-dilatation. Repeat arteriography at 11 mos shows no recurrence of RAS in case 1. No serious complications occurred.

We concluded that percutaneous transluminal angioplasty offers an acceptable alternative to surgery in renal transplant recipients with hypertension due to renal artery stenosis.

1521 HYPERSPLENISM(HS) IN RENAL TRANSPLANT RECIPIENTS(TR): THERAPY WITH PARTIAL SPLENIC EMBOLIZATION(PSE). Robert C. MacDonell Jr, Amil J. Gerlock, Jr, Winston C.V. Parris, H. Keith Johnson, Marion B. Tallent, Bruce I. Turner, Robert E. Richie, Gary D. Niblack (Spon. by Robert C. Boerth). Vanderbilt Univ. Medical Center; Depts. of Pediatrics, Medicine, Radiology, Anesthesiology, Surgery, Urology and Microbiology; Nashville, Tennessee.

HS complicates management of TR and limits immunosuppression, jeopardizing graft function. Splenectomy affords variable benefit but imposes risks of major surgery and potential infectious complications in immunosuppressed subjects. 3 TR and 1 patient awaiting transplantation developed severe leukopenia and/or thrombocytopenia and splenomegaly, not corrected by discontinuing possible offending drugs. 65-95% splenic infarction with Gelfoam particles were performed.

Case	Age (yr)	WBC ($1000/mm^3$)		PLATELETS ($1000/mm^3$)		IMMUNE SUPPRESSION (mg/day)		CREATININE (mg/dl)		F/U (mo)
		pre	*post*	*pre*	*post*	*pre*	*post*	*pre*	*post*	
1	20	1.8	10.4	179	258	12.5	62.5	5.0	3.4	3
2	18	5.8	4.2	51	109	0	100	1.8	1.9	5
3	16	2.4	4.1	171	364	12.5	100	1.7	1.6	2
4	22	2.0	12.1	143	400	- -	- -	6.5	7.1	1

In the TR improved hematologic parameters allowed increased immunosuppression and stable, or improved, graft function. Complications (fever, pain, ileus, hematoma) were not serious.

PSE offers significant advantages to these patients, avoids major surgery, and has proven effective in treatment of HS.