

## NEPHROLOGY

### 1045 CHANGES IN GLOMERULAR BASEMENT MEMBRANE (GBM) ANTI-GEN(S) WITH AGE AND DISEASE. Sudhir K. Anand, Benjamin H. Landing, Elin Lieberman, Eva Heuser, David Olson. Univ. So. Cal. Sch. Med., Child. Hosp. Los Angeles, Div. Nephrology and Dept. Pathology, Los Angeles.

GBM of patients with Alport Syndrome (AS) does not bind anti-GBM (McCoy et al, Lab Invest 3:19, 1976). Diagnostic use of staining kidney biopsies of AS suspect children with antiGBM was evaluated. Kidneys of 21 autopsied patients without renal disease (Groups I-III) and 3 patients with AS (with characteristic electron microscopic lesions) were treated with serum of a patient with strong antiGBM, then with fluorochromed antiIgG. Results were as follows:

GROUPS	PT. NO.	AGE RANGE	IMMUNOFLOUORESCENCE			
			0	1+	2+	or >
I	7	0 - 3 mo.	6	1	0	
II	5	3 mo. - 2 yr.	0	5	0	
III	9	3 - 30 yr.	0	1	8	
<u>p value for differences among Groups I, II &amp; III is &lt; 0.001</u>						
IV	3(AS)	7 - 15 yr.	3	0	0	

Young infants, like AS patients, appear to lack GBM antigen(s) normally present in older children and adults. Normal GBM increases from 950 Å at birth to 2844 Å by 3 years. Whether adult components are superimposed on fetal GBM or fetal GBM is replaced by adult GBM components is unknown. Failure of GBM to bind antiGBM is not diagnostic of AS in young infants, but is useful in older patients. These data may also partly explain absence of antiGBM nephritis in young children.

### 1046 MATURATIONAL CORRELATES OF NEPHRON STRUCTURE AND FUNCTION DURING DEVELOPMENT. Billy S. Arant, Jr. (Spon. by J. N. Etteldorf), University of Tennessee Center for the Health Sciences, Department of Pediatrics, Memphis.

Glomerulo-tubular (GT) relationships in the developing kidney have been characterized as immature, with anatomical and functional glomerular preponderance. Previous studies have not differentiated renal immaturity from other possible causes of GT imbalance. Glucose titration experiments were performed in 13 puppies from 2-51 days of age in which volume expansion (VX) was avoided (A), given 20ml/kg 0.9% saline IV (B) and observed (C). Proximal tubular length (L) and diameter of Bowman's capsule (D) were measured by microdissection studies. Maximal tubular reabsorption of glucose (TmG) and inulin clearance (Cin) increase with age (r=.96)\*, with dry kidney weight (DKW) (r=.72)\*\* and with L (r=.84)\*. DKW increased with L (r=.61)\*\*. Cin decreased with D (r=-.58)\*\*. Glucose threshold increased from 133 to 210mg/dl from 2-23 days (r=.84)\* but did not increase thereafter. The pattern of functional spay (S) < 3 weeks of age was greater than that observed > 3 weeks. TmG/Cin (3.37±.28,  $\bar{x}$ ±SD) did not change with age (r=.54). During B a decrease in TmG\*\* without change in Cin resulted in a decrease in TmG/Cin (2.97±.67)\*\* and an increase in S at all ages. During C, TmG increased, Cin did not change and TmG/Cin increased to 3.60±.99\*\*, values not different from A. S resembled that noted in A at saturation. It is concluded that increments in TmG, Cin and DKW during development, are related to nephron growth and that GT balance for glucose obtains from birth in the canine kidney whose response to VX is qualitatively similar to the adult at every stage of morphological development. \*p<.001, \*\*p<.05

### 1047 SERUM LIPIDS IN UREMIC CHILDREN AND THE RESPONSE TO NUTRITIONAL SUPPLEMENTATION. Watson C. Arnold, Maria G. Boosalis, Malcolm A. Holliday. University of California, Department of Pediatrics, San Francisco.

The incidence of hyperlipidemia and the effect of dietary factors on serum lipid values were evaluated in 17 non-nephrotic patients with GFR < 60 ml/min/1.73 m<sup>2</sup>. The mean serum cholesterol level (C) in 17 children was 214 ± 50 mg/dl and the mean serum triglyceride level (TG) in 12 children was 187 ± 67 mg/dl. C was elevated (> 230 mg/dl) in 22% and TG (> 140 mg/dl) in 92% of the children. There was no correlation between lipid levels and GFR, albumin, creatinine or BUN.

Multiple computer diet analyses were obtained before and during nutritional supplementation. Absolute values for calories and grams of protein, carbohydrate (CHO), and fat increased. However, when expressed as a percentage of calories ingested, CHO increased 11% while fat decreased 8% and protein remained unchanged. In 16 patients C increased from 207 ± 38 mg/dl to 239 ± 49 mg/dl (p < 0.025) and in 10 patients TG increased from 197 ± 66 mg/dl to 215 mg/dl (p < 0.1). C and TG were not related to total calories, gm/kg CHO or % fat ingested. TG was related to % CHO of total calories ingested. Diet appears to be one of the factors influencing elevated lipid levels in uremic children.

### 1048 RADIORECEPTOR ASSAY FOR SOMATOMEDIN-A IN UREMIC CHILDREN. Watson C. Arnold, E. Martin Spencer, Knut O. Uthne, Carolyn F. Piel, Malcolm A. Holliday. Univ. of Calif., Department of Pediatrics & Medicine, San Francisco.

The suggestion that the low Somatomedin A (SM-A) found in uremic children is due to the presence of a serum inhibitor to the bioassay was investigated using the human placenta radio-receptor assay. Twenty three children with chronic renal failure were divided into 3 groups based on the degree of renal failure.

Group	I	II	III
GFR (ml/min/1.73m <sup>2</sup> )	>50	<50	hemodialysis
SM-A U/ml & SEM	0.8±0.09	3.96±.58	9.92±2.6
No. of pts.	3	15	5

In the children in group II, there was no correlation between SM-A and GFR, BUN, creatinine, chronological age, bone age, or serum lipids. A correlation was found to growth expressed as a percentage of expected growth (r=0.59). A negative correlation was found to transferrin levels (p < 0.01). Six patients ingesting calories < 80% recommended dietary allowances (RDA) for height age had SM-A levels of 3.03 ± .68 U/ml. Nine patients ingesting > 80% RDA had values of 4.59 ± .81 U/ml (p < 0.05). In 3 patients paired SM-A levels rose with caloric supplementations from 3.22 U/ml to 5.1 U/ml (p<0.05). These findings demonstrate that: 1) SM-A levels are elevated in uremic children, 2) these levels correlate with growth, 3) nutritional status influences SM-A levels, 4) there may be an inhibitor in the serum of uremic children that interferes with bioassay.

### 1049 ANTICONVULSANT-INDUCED SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) AND NEPHROTIC SYNDROME (NS). Andrew J. Aronson, Hashallah Ezzati, Keyoumars Solteni and Rosa T. Ong (Spon. by Burton J. Grossman), The University of Chicago Pritzker School of Medicine, La Rabida Children's Hospital, Departments of Pediatrics and Medicine (Dermatology), Chicago, Illinois.

A 3 year old boy was treated with diphenylhydantoin (DPH), ethosuximide (E), and phenobarbital for convulsions following herpes encephalitis. After 3 months of anticonvulsant treatment he developed NS (24 hour urine protein-9.1 grams, serum albumin-0.76 gms.%, cholesterol-356 mg.%), positive ANIF (1:800), "suspectively" positive LE preparation, positive skin biopsy (dermal-epidermal IgG), and high total eosinophil count (985 - nl 50-250). After withdrawal of DPH and E there was gradual resolution of NS and normalization of urine, serum protein, cholesterol, ANIF and eosinophilia. Renal function and serum complement levels remained normal. Corticosteroids were not given.

Although anticonvulsants have been associated with SLE-like syndromes, serologic abnormalities, nephritis or NS, no previously reported patient developed SLE and NS concurrently. The urgency in discontinuing DPH and E prevented our identifying the offending drug. It is likely that this patient sustained a syndrome of drug-induced SLE with immune-mediated reversible glomerulonephritis and NS.

### 1050 RENAL HISTOPATHOLOGY AND CLINICAL COURSE OF 53 CHILDREN WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE). Andrew J. Aronson, Ronald J. Kallen, Burton J. Grossman, Phisit Saphyakhaion, Rosa T. Ong and Benjamin H. Spargo The University of Chicago Pritzker School of Medicine, La Rabida Children's Hospital, Departments of Pediatrics and Pathology, Chicago, Illinois.

53 patients with SLE have been followed for 7 months to 10 years and have undergone at least one renal biopsy (RB). Histopathologic classification of initial RB was: 1 normal, 9 Type I (proliferation without deposits), 5 Type II (membranous), and 38 Type III (proliferation with deposits). Each patient received prednisone, 13 received azathioprine or cyclophosphamide. Subsequent RB or post-mortem examination in 15 showed progression in 4 (1 with Type I, 2 with Type II and 1 with Type III), improvement in 4 with Type III and no significant change in 7. 4 developed renal failure, 3 of whom expired, 8 died of non-renal causes and 1 is azotemic. The remainder are alive. Chronic renal failure and persistent active renal disease occurred only with Types II and III. The low incidence of renal failure (7.5%) in this large series of patients with SLE despite the preponderance of Type III suggests a better prognosis than previously reported.