

Ever since the demonstration of the "swan-neck" lesion (shortening and narrowing of the neck of the proximal renal tubule) in cystinosis by nephron microdissection, it has been a source of speculation. The prenatal or postnatal onset of the lesion and its role in the pathogenesis of cystine storage have been debated. To study this problem, renal function tests, kidney biopsies, and renal tissue amino acid analyses were performed on two cystinotic infants at six and 12 months of age. At six months, the infants showed a generalized aminoaciduria, proteinuria and glucosuria but not hypophosphatemia, hypokalemia or polyuria; and they were pitressin responsive. Renal biopsies did not show the "swan-neck" lesion by light microscopy or nephron microdissection. However, electron microscopy revealed vacuolization and previously undescribed ultramicroscopic crystals in the epithelial cells of the neck region of the proximal tubule. The unbound cystine concentration in the kidney biopsies was three-fold greater than in controls. At 12 months of age the infants exhibited hypokalemia, hypophosphatemia, polyuria and subnormal pitressin responses. Typical "swan-neck" lesions were demonstrated by nephron microdissection. The unbound cystine concentration was ten times normal levels. These studies indicate the "swan-neck" lesion in cystinosis is an acquired rather than congenital defect and it follows rather than precedes cystine storage.

Paramedical personnel in evaluating children with renal disease. DEBBIE DEAN, BARBARA PETERS, and GEORGE A. RICHARD. *Univ. Fla. Coll. Med., Gainesville, Fla.* (Intr. by G. L. Schiebler).

During the past four years we have utilized the Clinical Laboratory Patient Service Assistant to help us evaluate children with renal disease. She arranges, conducts and calculates all studies, except for performing the renal biopsy. This has permitted us to evaluate 450 children with renal disease, including 2,000 complete series of renal function studies and 300 renal biopsies.

Previously, this diagnostic work-up required 5.5 hospital days. The Clinical Lab. Patient Service Assistant has permitted us to evaluate each patient in the outpatient dept. in 7 hours (overnight hospitalization required for renal biopsy). The studies include two 12-hour urine collections in the recumbent and ambulatory positions (done prior to coming to the hospital), concentrating and diluting capacity, urine culture and urinalysis, creatinine and urea clearance, tubular resorption of phosphate, blood gases, appropriate serological studies and blood chemistries, and a water load test. An intravenous pyelogram and voiding cystogram can be done before and after the studies are completed, respectively.

The Clinical Laboratory Patient Service Assistant has permitted us to decrease hospitalization time, decrease expenses to the family for lodging, decrease cost to the insurance carrier and the time of confinement in the hospital. She represents a familiar friend to the children with renal disease whom we follow.

Body composition in uremic children and the effects of chronic hemodialysis. CAROL J. WILSON, DONALD E. POTTER, JEAN L. HARRAH, MACARIO BUZON, and MALCOLM A. HOLLIDAY. *Univ. of Calif., San Francisco, and San Francisco Gen. Hosp., Calif.*

Chronically uremic children seldom grow normally; growth while on extended hemodialysis is usually not adequate. In order to delineate more precisely the components of this growth failure and the changes in body composition with chronic

dialysis, 46 simultaneous measurements of total body water and of extracellular water (ECW) were determined in 24 children aged 2-17 yrs. The data were divided into two groups: uremic children dialyzed <3 mos and those dialyzed >6 mos. Published data of Cheek for healthy children of similar age were used for comparison. In the uremic children body weight (BW) and height (Ht) were below normal for age. ECW was significantly higher in both groups for Ht and BW, and its fluctuations tended to confuse interpretation of weight changes. In both groups, cell mass (CM), derived from calculated intracellular water, was low in relation to BW and Ht. While on dialysis, Ht, BW, and CM tended to increase; CM as per cent BW did not change, and increases in CM with Ht were less rapid than in normals. Correlation between changes in CM with change in Ht was low. Nutritional status was a factor in rate of increase in CM and in Ht. Although normal linear growth velocity may be observed on dialysis, catch-up growth is uncommon. Growth occurred without restoration of the normal relationship of CM to Ht. These studies have proved useful in defining the character of body composition in uremic children and in evaluating changes in body composition in individual patients with time, variations in caloric intake, and with growth.

#### GASTROENTEROLOGY AND ENZYMOLOGY

Growth of the small intestine in IUGR and normal rat pups. M. K. YOUNOSZAI (Intr. by Samuel J. Fomon). *Univ. of Iowa Coll. of Med., Univ. Hosp., Iowa City, Iowa.*

Small intestinal growth was compared in intra-uterine growth-retarded (IUGR) pups, obtained from rats fed a diet containing 6% protein during pregnancy and normal (N) control pups. All pups were delivered by C-section at term and nursed with normal foster mothers (7-9 pups/litter). At birth mean body weight in IUGR (27 pups) was 4.3g and in normals (28 pups) 6.3g. Mean small intestinal weight (mg) in IUGR pups was less than in normals (IUGR = 105, N = 140,  $p < 0.01$ ). The intestinal length (cm) was also shorter in the IUGR pups (IUGR = 19.7, N = 21.1,  $p < 0.01$ ). The intestinal hypotrophy (relative low weight/cm) in IUGR pups was more marked in its distal one-third than in the more proximal segments. By age 8 days body weight (16g) and intestinal weight (470mg) were similar in the two groups. However, in the IUGR group hypotrophy was still noted in the distal one-third of the intestine. At this age intestinal length was greater in IUGR pups than in normals (IUGR = 36.4, N = 34.1,  $p < 0.05$ ). Intestinal tissue water and protein contents were similar in IUGR pups and normals. However, protein content (% wet wt.) in the middle one third of the intestine was higher at 8 days than at birth (IUGR, N: 0 days = 14.5, 14.6, 8 days = 16.8, 16.2;  $p < 0.01$ ).

Thus the relatively more rapid body growth in IUGR pups was associated with a similar rapid growth of the small intestine which was more prominent in the proximal than in the distal segments. The higher protein content in the mid-gut at 8 days compared to that at birth suggests a better functional capability of this segment at 8 days than at birth.

Mechanism of antigen absorption from the small intestine. W. ALLAN WALKER, RICHARD CORNELL, LAURA M. DAVENPORT and KURT J. ISSELBACHER. *Harvard Med. Sch., Mass. Gen. Hosp., Boston, Mass.* (Intr. by J. Warshaw).

Evidence exists that the intestine is permeable to antigenic macromolecules. The mechanism of absorption and factors af-