Pediat. Res. 3:364, 1969

70 *Accumulation of Cystine Following Renal Homotransplantation for Cystinosis.* K. MICHAEL HAMBIDGE*, STEPHEN I. GOODMAN*, PHILIPPE A. WALRAVENS*, S. MICHAEL MAUER*, LAWRENCE BRETTSCHNEIDER*, ISRAEL PENN* and THOMAS E. STARZL*, Departments of Pediatrics and Surgery, University of Colorado School of Medicine, Denver, Col. (introduced by Donough O'Brien).

Cystinosis is characterized by high intracellular and normal plasma levels of cystine. There is experimental evidence to indicate that the intracellular accumulation derives from a primary defect in cysteine-cystine metabolism in the affected cells. It is expected, therefore, that renal homotransplantation in cystinotic children would not be complicated by accumulation of cystine in the normal cells of the donor kidney.

Bilateral nephrectomy and cadaveric renal homotransplantation was performed on a 10-year-old cystinotic male (renal cystine = 58.1 μMol/g wet weight). Five months later creatinine, phosphorus, and amino acid clearances were normal, although cystine had accumulated to 2.9 μMol/g wet weight, the highest tissue content of any one free amino acid. Renal cystine content of 5 non-cystinotics and one obligate carrier was less than 0.5 μMol/g wet weight.

In these circumstances, it would appear that additional extracellular factors must be involved in the abnormal cystine deposition in this disorder. (SPR)