CYCLIC AMP AND SOME RENAL TUBULAR DYSFUNCTION SYNDROMES. M. Vanderschueren-Lodeweyckx**, E. Eggermont, R. Eeckels, L. Corbeel*, W. Proesmans
Cyclic adenosine 3',5'-monophosphate (c-AMP) and its dibutyryl derivative (DBc-AMP) were given intravenously, either as a single shot in two different dosages (c-AMP: 6 and 10 mg/kg, DBc-AMP 5 mg/kg) or as an infusion for 45 to 60 minutes (c-AMP: 0.5 mg/kg/min, DBc-AMP: 0.2 mg/kg/min) to six patients with nephrogenic diabetes insipidus (NDI), to one boy with cystinosis and to one girl with Bartter's syndrome. c-AMP as a single shot or as an infusion was unable to reduce, in a constant and reproducible manner, urine volume (V) and free water clearance (C_{H_2O}) in NDI patients and in the cystinosis patient. DBc-AMP given to the same patients regularly did increase these parameters. In contrast to this, in the patient with Bartter's syndrome, both c-AMP and DBc-AMP reduced V and C_{H_2O} significantly.

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ALPHA₁ ANTITRYPSIN (α_1 AT) AND LIVER DISEASE.

A. Sass-Kortsak, S.P. Moroz, J.F.T. Glasgow, S. Graham, D.W. Cox, E. Cutz. Dep. of Pediatrics, University of Toronto, and Research Institute, Hospital for Sick Children, Canada. Observations on 16 children with liver disease, α_1 AT deficiency and Pi (protease inhibitor) type ZZ(14) or SZ(2) will be presented. Of 18 sibs of 12 ZZ probands, 6 were of type ZZ, 2 affected and 4 unaffected. Only 13 of the 16 patients presented with early jaundice and "neonatal hepatitis". Two were never jaundiced (both ZZ) and one only at 2 y of age (SZ). Two sibs and one additional patient (all Pi ZZ) had both liver and lung involvement. Three of the 16 patients died of liver failure at 5, 10, 12 y of age. Eleven patients are alive, 7 presently under 5 y of age, 8 between 5-15 y. Of these, only 2 appear severely involved, 2 moderately sick and 7 are relatively well and only mildly affected. Prognosis is, however, uncertain in all. Careful search for possible other associated etiological factors (Au antigen and antibody, CMV, rubella, toxoplasmosis, lues, cystic fibrosis, etc.) was negative except for renal tubular ectasia and bladder neck obstruction in one each. One patient with severe "neonatal hepatitis" in infancy and one with "chronic active hepatitis" at 7 y of age were found to be Pi type MZ. In view of our group having found abnormal lung function in Pi MZ individuals the possible etiological significance of Pi type MZ (with intermediate deficiency of α_1 AT) in liver disease is entertained but is regarded as far from proven.