55 HYDROGEN ION EXCRETION IN NEPHROTIC SYNDROME IN CHILDREN

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H^+_{NAE} excretion was studied in 3o children with oedemas /age of 2-1o yrs/. Plasma creatinine, urea nitrogen, Na were normal, GFR was slightly decreased. Blood acid-base equilibrium was shifted towards metabolic acidosis. Mean H^+_{NAE} excretion was significantly decreased before and after NH_4 Cl loading. Before loading it was 35,75 uEq/min/1,73 m², after loading in 3 and 5 hours 67,14 uEq/min/1,73 m², 78,17 uEq/min/1, 73 m²/respectively. Decrease in H^+_{NH4} excretion was more significantly expressed then H^+_{TA} excretion. Drop in H^+_{NAE} excretion was positively correlated with diuresis, GFR, and tubular reabsorption of Na. Negative correlation between plasma t CO_2 and H^+_{NAE} excretion was not observed in contrast to the healthy children. Children with nephrotic syndrom showed lower ability for H^+_{NAE} excretion before and after NH_4Cl loading.

56 BONE AGE IN TWINS

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71 twin pairs, aged 8-12 yrs, have been investigated. Zygosity has been determined by eight blood group systems. 25 pairs/14 female, 11 male/were monozygous, 19 pairs/ 1o female, 9 male/were like-sexed dizygous, 27 pairs unlike-sexed. Bone age was measured by the Tanner-Whitehouse method. Twins as a group are not retarded in skeletal maturation at this age, although 34 % were born prematurely. Mean difference/ intrapair/ in bone age was o.o96 ± o.o73 /SD/, o.918 ± o.616 andl. 149 ± o.58lyrs for the 3 groups respectively. P<o.ool for MZ/DZ, P=o.5 for DZ like-sexed/ unlike-sexed. Within-pair correlation coefficiens are o.996 /MZ/, o.735 /DZLS/ and o.517/DZUS/. Bone maturation at this age is determined largely by genetic factors.

57 A NEW AND NON INVASIVE METHOD FOR DETERMINING BODY DENSITY IN NEWBORN BABIES

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A method will be described of measuring body volume which depends on placing a baby for a few seconds in a closed perspex box and injecting a known quantity of air into the box. The pressure change enables one to calculate the volume change in the air of the box and thus the baby's volume. This principle is well known but is too insensitive for accurate results. Accurate determination has been made possible by employing a second box of identical dimensions, a differential pressure transducer between the two, and cyclical pressure changes to eliminate artefact caused by the baby's breathing and moving. Total body fat measurements in small for dates, and in preterm babies are given.

58 DIAZEPAM KINETICS IN INFANTS WITH COELIAC DISEASE

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Whenever gastrointestinal function is impaired, the clinical problem of drug absorption and disposition has to be considered, if a rational therapeutic scheme has to be followed. In four infants with a very severe form of coeliac disease a complete analysis of diazepam /DZ/ kinetic was performed, both during the active stage of the disease and after a prolonged period of gluten free diet. With the informed consent of the parents, serial blood samples were drawn during the first 24 hours after single oral administration of o.3 mg/kg b.w. of DZ, to measure DZ and demethyl-DZ plasma concentration. Urine were collected for 72 hours, to determine free and conjugated metabolites excretion. Much higher peak concentration values of DZ were observed during active stage of the disease together with a smaller volume of distribution. No significant variations of drug absorption were noticed. The relative importance of deep variations of body composition in malnourished coeliac infants and of impaired rate of drug metabolism will be evalutated.

59 ALFA-FETOPROTEIN /AFP/, a_1-ANTITRYPSIN ACTIVITY AND HBsAg IN NEONATAL HEPATITIS AND BILIARY ATRESIA

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AFP, a_1-antitrypsin activity, and HBsAg were studied in 29 young infants, with prolonged direct reacting bilirubinemia; 4 had biliary atresia, and 25 neonatal hepatitis syndrome, due mainly to type B hepatitis /13 cases/. A high death rate /5 out 25/ during the acute phase, was related to type B hepatitis. AFP was not detected in patients with biliary atresia, but was increased in 22 /88%/ of patients with neonatal hepatitis. All 3 patients with negative AFP had a fulminating type B neonatal hepatitis and died in hepatic coma. One of them had also a_1-antitrypsin deficiency. All others had normal a_1-antitrypsin. These results suggest that the prognosis of neonatal hepatitis is grave in patients with type B hepatitis associated with AFP and a_1-antitrypsin deficiency.

60 ORNITHINE TRANSCARBAMYLASE DEFICIENCY IN REYE'S SYNDROME

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Hyperammonemia is observed regulary in patients with Reye's Syndrome /RS/, and may be a crucial determinant of the encephalopathy in this often lethal condition. Since ammonia detoxification depends on an intact urea cycle and since patients with inherited defects of a single urea cycle enzyme also demonstrate hyperammonemia, we sought to define the mechanism of hyperammonemia in RS by examining activities of the first two urea cycle enzymes, carbamyl phosphate synthetase I /CPS I/ and ornithine transcarbamylase /OTC/ in liver homogenates from 4 RS patients. OTC activity was reduced to 1o-15 % of control mean in 2 autopsy specimens and to 4o-42 % of control in 2 biopsy samples. In 2 livers K_m's for the substrates, ornithine /Orn/ and carbamyl phosphate /Cp/, were normal and the V_{max}'s were much reduced; in a third, the K_m's for Orn and Cp were increased 1o fold and 5 fold, respectively, but the V_{max}'s were normal. Significantly CPS I activity was normal in all 4 livers and activity of a third urea cycle enzyme, arginase, was normal in the one RS liver assayed. These data suggest that specific deficiency of OTC activity may be responsible for the hyperammonemia in RS and that more then one kind of kinetic disturbance may underlie the OTC impairment.