

Cardiovascular Risk Factor Status of Greek Adolescents in Athens

As it has been well documented recently, the early recognition in childhood and adolescence of risk factors for catastrophic conditions in the adult life, such as cardiovascular diseases and cancer, could be the only effective preventive measure for these diseases if these persons had an appropriate management. In order to establish the frequency of such risk factors in adolescents aged 10-15 years a collaborative study was carried out in 11 countries with the initiative of the American Health Foundation. A total of 1,113 Greek adolescents of both sexes aged 10-15 years attending public schools of the area of Athens was examined. For each adolescent a self-administered questionnaire was filled, blood pressure, pulse rate, anthropometric measurements taken and plasma cholesterol was estimated. One or more risk factors (hypertension, hypercholesterolemia, smoking, obesity - weight over the 90th percentile) were present in 34% of the Greek adolescents, who also had the highest mean quetelex index (weight to height square) among the 11 participating countries. The mean systolic and diastolic blood pressure were among the highest while the average plasma cholesterol (151.4mg/dl) was the lowest among the 11 countries. Although Greece has still a low cardiovascular disease mortality in comparison to other countries, the high incidence of risk factors in the adolescents found in the present study, may indicate an increase of morbidity and mortality in future generations.

Atherosclerosis Precursors in Finnish Children. Results of A Pilot Study.

The objective of the present multicenter study was to obtain information on the risk factor levels for coronary heart disease and their determinants in children of various ages in different parts of Finland, and to provide facts for the planning of intervention. The pilot study, carried out in 1979 comprised 3-, 12-, and 17-year old subjects. 520 children were selected by random sampling; 71 % participated. Height, weight, skinfolds, blood pressure, and biochemical parameters (serum cholesterol, HDL-cholesterol, triglycerides, immunoreactive insulin, urinary NaCl) were measured, a dietary survey (48 hour recall) was carried out, and attention was also paid to psychosocial factors. Mean serum cholesterol in 3- and 12-year old children was 4.9 mmol/l, which is high by international standards (Knuiman et al.: Lancet 1979;II:1183), and HDL-cholesterol was 1.4 mmol/l. 14 %, 37 % and 49 % of the total energy intake was derived from protein, fat and carbohydrate, respectively. The P/S ratio in the diet was lowest, 0.18 in the north, and highest, 0.24 in the Helsinki region. - The data serve as a background for a larger cross-sectional study, to be carried out in 1980.

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Neonatal Hypoxic-Ischemic Encephalopathy: clinico-radiological(CT) correlations in acute and chronic state.

The role of Computed Tomography(CT) after perinatal asphyxia in term infants was studied prospectively in 28 infants and retrospectively in 8. 16 CT were obtained during the acute state("early", day 1-7), 19 during clinical recovery("intermediate", week 1-4), and 16 in chronic state("late">3 months). In early CT obliteration of ventricles or cisterns was not a constant sign of clinically suspected generalized brain edema. Hypodensities in the CT were taken as indicators of local edema or necrosis. In early CT their extension and degree were of little diagnostic value, but in intermediate CT they correlated well with neonatal neurological signs (i.e. status epilepticus, contralateral hemiconvulsions), and as far as follow up (8-30 months) is known, were of good prognostic value. In late CT, hypodensities are replaced by ventricular dilatation (with or without cortical atrophy) usually generalized and symmetrical, rarely very localised, or disappeared leaving no detectable pathology. Findings in late CT correlate poorly with clinical sequelae. Thus intermediate CT seems optimal for evaluation and prognosis of hypoxic-ischemic brain damage. (Suppt.by Kamillo-Eisner St.and Swiss Nat.Found)

Hacettepe Univ. Children's Hosp. Medical Center, Pediatric Hematology Unit, Ankara, Turkey and Claude Bernard Univ. Dept. of Molecular Biology, Lyon, France.

Molecular Hybridisation Studies in Beta<sup>0</sup> Thalassemia

The beta thalassemias are heterogenous disorders at the molecular level. In order to understand the molecular pathology of beta<sup>0</sup> thalassemia, the hybridisation was done by using DNA and complementary DNA(cDNA). The patient studied was homozygous for beta thalassemia. Whole cell incubation in the presence of <sup>3</sup>H-L-Leucine and separation of globin chains were performed as described by Clegg et al. Normal human globin mRNA was isolated from the total RNA of reticulocytes. cDNA alpha and beta was prepared using reverse transcriptase with <sup>3</sup>H-d-CTP. Normal DNA was prepared from a normal placenta and beta<sup>0</sup> thalassemia DNA was isolated from the spleen of our case. Normal DNA and beta<sup>0</sup> thalassemia DNA were mixed with increasing amounts of cDNA. The percentage of cDNA hybridized was converted to picogram cDNA. This study showed that there was no difference between normal control and beta<sup>0</sup> thalassemia patient according to the hybridisation with DNA and cDNA. This finding suggests the absence of deletion in our patient.

Globin synthesis in beta-thalassemia major without anemia. A 2½ year old girl (N.B.) with homozygous beta-thalassemia (thal.) with only mild anemia (Hb 10.6-12.8 g/dl) and with complete absence of HbA, which is replaced by 1.3 % HbA<sub>2</sub> and the rest being HbF, is presented. Her parents show both heterozygous beta-thal. with increased HbA<sub>2</sub> (5.5-5.9%) and HbF (6.0-6.9%). The disease of N.B. differs from homozygous forms of beta<sup>0</sup>-thal. by the absence of severe anemia, of beta<sup>+</sup>-thal. by the lack of HbA and from beta-thal. or hereditary persistence of HbF (HPFH) by the presence of HbA<sub>2</sub>. Her beta-thal. is similar to the one reported in two homozygous individuals of a dutch sib by Schokker et al (Nature 209:44,1966). Incorporation of <sup>3</sup>H-leucine in reticulocytes and separation of the globin-chains confirms the absence of beta-chain-synthesis in N.B. and the non alpha:alpha-chain synthetic ratios are unbalanced for N.B. (0.47) which is distinctly above the ratios reported for homozygous beta<sup>0</sup>-thal. (0-0.2). Absence of severe anemia in homozygous beta<sup>0</sup>-thal. and HPFH was recently shown to be related to the absence of the delta-gene and its flanking DNA-sequences. Homozygous beta<sup>0</sup>-thal. and beta<sup>+</sup>-thal. was thus proposed to lead to severe anemia by the presence of a regulatory DNA-sequence near the delta-chain locus suppressing gamma-chain-synthesis, which in beta<sup>0</sup>-thal. and HPFH replaces the lacking beta-chains (A. Banks, Science 207:486, 1980). In N.B. the gamma-chain-suppressor seems to be inactive as in beta<sup>0</sup>-thal. but the delta-gene is present, thus indicating the separation of the gamma-regulatory gene from the delta-structural gene.

REDUCED CATALYTIC ACTIVITY FOR GLYCOGEN OF ACID a-GLUCOSIDASE TYPE 2 FIBROBLASTS.

The lysosomal enzyme acid a-glucosidase (a-G) catalyzes the cleavage of glucose from glycogen. Three phenotypes of a-G, 1,2 and 2-1, determined by two alleles at an autosomal locus, have been identified in normal subjects (Ann. Hum. Genet., 38:391, 1975).

Using maltose as substrate the a-G activity in cultured fibroblasts (FB) homozygous for a-G type 1 (common allele) was 233 ± 95.1 µg glucose/mg protein/hr, whereas the activity in a FB strain homozygous for a-G typw 2 was 81.5 (35% of normal). Using glycogen as substrate the activity in the normal fibroblasts was 124.8 (53.4% of that obtained with maltose), whereas in the a-G 2 FB it was 8.4 (10.3%). The specific activity, therefore, in the a-G 2 FB when glycogen was the substrate was only 6.7% of that measured in the a-G 1 FB. By using the same amount of enzyme activity (measured with maltose) the size of the precipitin arcs obtained by rocket immunoelectrophoresis was the same in a-G types 1 and 2 FB. These findings indicate that FB homozygous for a-G 2 are deficient toward glycogen because of a functionally abnormal enzyme. Since the residual activity in the FB of patients with the adult form of a-G deficiency may be up to 22% of normal, it is possible that homozygotes for a-G 2 may develop a muscular dystrophy-like disease later in life.