A NEW VARIANT OF GLUCOSEPHOSPHATE ISOMERASE (GPI) DEFICIENCY WITH CONGENITAL HEMOLYTIC ANEMIA.
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A new variant of red cell GPI deficiency (Type Nordhorn) with congenital nonspherocytic hemolytic anemia is described. The propositus suffers since birth from a severe anemia. GPI activity is decreased to 22% of the normal. The parents exhibit activities between 36 and 47 %. The thermal stability of the mutant enzyme is decreased in the propositus and in all affected maternal relatives. The propositus is not homozygous, but double-heterozygous for two abnormal alleles. The heterozygous mother contributes an allele which produces a thermolabile enzyme of decreased activity and abnormal electrophoretic mobility, whereas the father contributes an inactive gene product. The enzyme defect becomes also manifest in the leucocytes of the propositus. The thermolability is also evident in the leucolysates of the mother. Compared with erythrocyte populations of similar mean cell age the rate of red cell glycolysis is reduced in the propositus. A premature inactivation of the enzyme during maturation of the erythrocytes is suggested.

HYPOTHYROIDIE PAR DEFAUT CONGENITAL DE SYNTHESE DE LA THYROGLOBULINE. **Ch. Sultan, R. Jean, S. Lissitzky, M. Michel-Bechet, J. Bismuth (Montpellier, France**

Chez 3 enfants dont 2 frères atteints d'hypothyroïdie par trouble de l'hormonosynthèse avec test au perchlorate de potassium et de charge en DIT normal, l'administration de 50 micro-curies d'Iode 135 permet d'obtenir les renseignements suivants: -Dans le sérum l'iode marqué est fixé sur les protéines anormales dénuées d'activité hormonale. -Dans le tissu thyroïdien prélevé par biopsie: . Quantité de protéines en mg/g de tissu = 96-102-46,8 = Normal. .Iode stable dans les protéines=0,oo2%-0,o67%-0.38%=élevé. .Après ultracentrifugation en gradient de saccharose=absence de radio-activité et de densité optique dans les zones 19 S et 12 S de la thyroglobuline, présence d'un pic unique. .Dosage radio-immunologique= absence de thyroglobuline. .Immuno-électrophorèse=un seul arc avec sérum antialbumine. .Auto-radiographie=seul l'arc albumine apparait radioactif.--Etude histologique= micro-adénomes microfolliculaires.

Conclusion: Absence de synthèse de la thyroglobuline remplacée par l'albumine dont l'iodation ne permet qu'une faible formation d'hormone.

ENDOCRINOLOGY

THE URINARY 11-OXYGENATION INDEX DURING CHILDHOOD+ **W.M.Teller, S.B.Pal, and J.Homoki. Dept. Pediatrics, Endocrinology and Metabolism. Univ. of Ulm/Donau, Fed. Rep. Germany**

The urinary 11-oxygenation index (O.I.) is the ratio of the quantity of corticosteroids not oxygenated at C 11, such as pregnanetriol, tetrahydro-11-dehydrocortisol etc. (=abnormal metabolites) to that of corticosteroids oxygenated at C 11 like tetrahydrocortisone, tetrahydrocortisol etc. (=normal metabolites). O.I. was examined in 5 groups of normal persons of various ages and both sexes (total number n=111): I:49 newborns, aged 0-4 wks., II: 17 infants, aged 1-12 mo., III: 16 small children, aged 1-6 yrs., IV: 19 school children, aged 7-13 yrs., V:10 adolescents and adults, aged above 13 years. The procedure for determination of O.I. was followed as described by Edwards et al. (J.Endocr. 30:181, 1964). The results were as follows (mean ± 2 S.D.): I: o.22 ± o.6, II: o.2o ± o.48, III: o.19 ± o.64, IV: o.17 ± o.4o, V: o.21 ± o.44. There was no sex difference. For the first time it was shown that irrespective of age O.I. remains the same throughout childhood (including the early newborn period). This property of O.I. provides the basis for its general usefulness in rapid diagnosis and surveillance of children with congenital adrenal hyperplasia.

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STUDY OF THE CONCENTRATION OF THYROID HORMONES IN THE AMNIOTIC FLUID. R. Tojo, J.L. Iglesias Diz, H. Iglesias. University Clinics of Pediatrics, Gynecology, and Pharmacology, University of Santiago de Compostela, Spain.

The objective of our study is to appraise the level of thyroid hormones in the amniotic fluid during pregnancy in women with goiter and/or hypothyroidism in a geographic region of endemic goiter (Galicia, northwestern Spain).
The parameters studied were: Index of T3, T4 and index of free thyroxin (FTI).
The determinations were performed in the amniotic fluid at the seventh month of pregnancy, and during childbirth, in women with goiter, and in the blood of the cord of their children. The results were contrasted with those obtained simultaneously in a group of pregnant women normal from the thyroid point of view and in their children.
The values obtained and the relationships of maternal-fetal thyroid function through the amniotic fluid, as well as the implications with regard to a possible prenatal preventive program, will be fully discussed at the meeting.

GASTROENTEROLOGY

GASTRIC LIPOLYSIS OF HUMAN MILK LIPIDS IN INFANTS WITH PYLORIC STENOSIS.
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In human infants and puppets a suboptimal concentration of bile acids in the duodenum may interfere with the micelle formation and so the absorption of dietary glycerides. In 1970 Helander and Olivecrona described, in the suckling rat, a lipolytic activity in the stomach. Such an activity has also been found in humans but is thought to be of quantatively minor importance in the normal fat digestion in the adult. We have found in infants a significant hydrolysis of milk triglycerides into diglycerides and free fatty acids to occur already in the stomach. The low pH, the relatively high content of diglycerides in the gastric samples and the nature of the disease of these infants suggests that the hydrolysis was not catalyzed by pancreatic enzymes. The pregastric lipase activity can be of great importance in the postnatal digestion of milk triglycerides in the human infant and to some extent compensate for the poor bile acid concentration found in these infants.

PEPTIDASE ACTIVITIES OF THE BRUSH BORDER OF RABBIT SMALL INTESTINAL MUCOSA. **G.Andria, A.Marzi and S.Auricchio. Dept. of Pediatrics, II School of Med., Univ. Naples, Italy.**

The peptidases located in the intestinal brush border (BB) probably play a major role in the pathophysiology of protein digestion, but their properties are still incompletely known. In the present study, rabbit BB activities hydrolyzing Phe-Ala(PA), Leu-Gly-Gly (LGG) and Leu-β-naphthylamide (LNA) showed high specific activities and amounted to about 20%, 15% and 50%, respectively, of the total mucosal activity for the same substrate. PA hydrolase of the BB differed from that of the cytosol on the basis of the following data... (mean ± SEM):

Puromycin (1mM)	**(BB) 51.2±1.5**	**(cyt) 95.5 3.5)**
pOH-MercuriBenzoate (0.1mM)	**101.5±0.8**	**10.3±0.4)**
Heat (50°, 60 min)	**68.2±10.2**	**11.3±5.0)**

Ion exchange chromatography separated the BB peptidases, after solubilization with papain, in 3 peaks. The main peak was active towards dipeptides (PA, Gly-Leu, Lys-Leu), tripeptides (LGG, Phe3) and arylamides (LNA, Lys-NA), whereas a 2nd one hydrolyzed dipeptides, with no LNAase and very low tripeptidase activity. PA hydrolases of both peaks were phMB and heat insensitive and only that of the 1st peak was inhibited (about 50%) by puromycin. The 3rd peak contained the γ-Glu-NAase activity.