101

POSTNATAL ENDOGENOUS GLUCOSE PRODUCTION RATE (EGPR) RELATED TO SEVERITY OF INTRA-UTERINE GROWTH RETARDATION (IUGR).

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The increased risk to hypoglycaemia in IUGR newborn infants is thought to be due to a limited EGPR. One might speculate that EGPR will be more reduced in infants with more severe IUGR. We measured EGPR in 16 IUGR infants with the prime dose-constant rate infusion technique using 6,6D₂ glucose. Gestational age ranged from 30 to 40 weeks (35±3 wk, mean±SD), birthweight ranged from 0.9 to 2.2 kg (1.7±0.4 kg). The infants were studied at a mean postnatal age of 9 hours. The infants received a low dose glucose infusion (mean 3.5 $mg/kg.min^{-1}$).

Results: Despite the low dose glucose infusion EGPR ranged from 0.4 to 3.2 $mg/kg.min^{-1}$. EGPR increased significantly with increasing gestational age (r=0.55, P=0.03) and birthweight (r=0.55, P=0.03). If the degree of IUGR was expressed in Standard Deviation Score (SDS) and Brain/Body Ratio (BB-ratio) we found no significant correlation between EGPR and SDS. However, with increasing BB-ratio, EGPR was significantly (r=-0.54, P=0.03) lower.

Conclusion: The lowest EGPR was measured in infants of shortest gestation and with the lowest birhtweight. IUGR infants with an asymmetrical growth retardation, who thus have a high BB-ratio, seem to have an increased risk to develop hypoglycaemia, since they have the lowest EGPR.

102

HEREDITARY ELLIPTOCYTOSIS WITH INFANTILE POIKILOCYTOSIS: TEMPORARY SEVERE HEMOLYTIC ANEMIA DURING THE NEONATAL PERIOD DUE TO SPECTRIN α-CHAIN VARIANTS.

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Despite its frequency the clinical importance of hereditary elliptocytosis (HE) is rather low, since most carriers of the disease never develop any clinical symptoms. Patients with clinical signs of HE mostly show a moderate to severe poikilocytic hemolytic anemia only during the neonatal period (HE with infantile poikilocytosis). Until now several hereditary defects of the red cell membrane skeleton have been recognized as cause of HE with infantile poikilocytosis. In normal erythrocytes the predominant membrane protein, spectrin, selfassociates to tetrameric molecules built of two α- and two ß-chains. We describe a diminished polymerization of spectrin due to two different defects of the α-chain in two unrelated German families with HE. The affected members showed a severe autosomal dominant inherited poikilocytic hemolytic anemia in the neonatal period. Afterwards a clinically inapparent elliptocytosis persisted. On agarose gel electrophoresis the concentration of unpolymerized dimeric spectrin was elevated to 17 - 50 % (normal < 10 %).

In one of the families the biochemical defect could be identified by limited tryptic digestion of spectrin. There was an elevation of an abnormal 74000 dalton peptide instead of the normal 80000 dalton peptide known to derive from the digestion of the α I domain. The association constant for the tetramer α'ß αß was determined as 0.5 x 10^6/M (normal 1.1 x 10^6/M). In the other family we found a splitting of the α chain into two bands on SDS polyacrylamide gel electrophoresis: the normal α chain (MW 240000) and an abnormal α' chain (MW 230000). The amount of the α' chain was increased after incubation indicating an increased susceptibility to proteolytic degradation.

The decreased spectrin polymerization causes an instability of the erythrocyte membrane with formation of elliptocytes. In the neonatal period the membrane stability is further diminished by the increased level of free 2,3 DPG, leading to severe poikilocytic anemia.

103

IVGG TREATMENT OF KAWASAKI SYNDROME (KD).

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We compared a single large dose of intravenous gamma globulin (IVGG) with the standard regimen of four smaller daily doses in reducing systemic inflammation and coronary artery abnormalities in 548 patients (pts) with acute KD within the first 10 days of fever in 7 U.S. university medical centers. Pts were randomly assigned either to a single-infusion (273 pts) (IVGG, 2 g/kg, over 10 hours), or to four-infusion (275 pts) (400 mg/kg/day for 4 consecutive days); both received aspirin, 100 mg/kg/day, through illness day 14, then 3 to 5 mg/kg/day. The groups were similar at enrollment. In the single-infusion group, mean decrease in temperature from study day 1-2 was greater (p<.0001) and albumin (p=.02) and α-1-antitrypsin (p=.05) improved more rapidly. Echocardiograms interpreted blindly and independently by two readers showed coronary artery abnormalities on one or more studies in 6.4% of pts overall: single-infusion vs four-infusion = 4.6% vs 8.2%, respectively, including those with abnormal echocardiograms at enrollment and 3.4% vs 5.5% excluding those with initial abnormalities. Side effects were equal and mild. Single infusion IVGG is safe and effective, reducing fever, inflammation and coronary artery abnormalities more than the 4 day regimen.

104

GRANULOCTYE-, COMPLEMENT- AND LYMPHOCYTE-FUNCTIONS IN HIV-INFECTED CHILDREN

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The spectrum of opportunistic infections seen in HIV disease in children resembles that in adults. Most striking, however, is the role of bacterial infections in pediatric AIDS. Although defective humoral immunity seems to play a significant role in predisposing to bacterial infections, there is no direct correlation between the risk for bacterial infections and poor B cell mitogenic responses or hypergammaglobulinemia (Rubinstein 85), suggesting additional predisposing factors.

We therefore examined polymorphonuclear leukocytes (PMNL) functions and complement activity (CH 50) in 17 HIV-infected children (P2A-D) in comparison with 37 children born to HIV infected mothers (P0). Lymphocyte subsets and functions (PWM and tetanus toxoid [TT] induced proliferation) as well as global and specific [TT-Ab] IgG levels were studied in parallel.

Our results confirm the well known quantitative (CD 3, HLA-DR normal; CD 4 decreased; CD 8, act. T-cells increased) and functional (PWM, TT induced stimulation decreased; IgG increased; TT-Ab decreased) disturbances of lymphocytes in HIV-infected children. In addition, PMNL of the P2A-D group show a reduced chemiluminescence to zymosan (p<0,05). No significant differences were observed in PMA induced chemiluminescence or CH 50 in the two groups.

From our data we conclude, that in HIV-infected children a reduced PMNL responsivness might contribute to a increased risk for bacterial infections.

105

REDUCED SIZE ORTHOPTIC LIVER TRANSPLANTATION (RSOLT) IN FINNISH CHILDREN.

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After 1987 10 RSOLT and one OLT have been performed in 10 pts at a mean age of 4.3y (range 0.6-14.2); in 3 pts over the ABO barrier.The diagnoses were: biliary atresia (4 pts),tyrosinemia (3 pts),alpha-1-antitrypsin deficiency,liver adenoma and hepatoblastoma.Triple immunosupression (MP,Az and CyA) with constant i.v. CyA infusion (B-CyA-spes=500-600 ug/L) for 2 w and MP+CyA after 2 mo.Two arterial thromboses were seen; one led to re-OLT and the other cleared with local i.v. streptokinase infusion for 2d.Five pts had one rejection 5-21d (mean 12) after OLT and responded to 3 mg/kg/d p.o. MP for 5d.There was no rejection in the 3 pts were the ABO barrier was crossed.CMV viremia with clinical symptoms was seen in 5 pts and treated with gancyclovir.

After a mean follow-up time of 0.8y (range 0.3-2.5y) all pts are at home with a functioning graft.Psychomotor development and growth (catch-up growth after 6 mo) are normal, mean SGPT is 30 U/L, gamma GTP 26 U/L and creatinine 49 umol/L.

106

PREVALENCE OF ANTI-HIV ANTIBODIES (HIV-Ab) IN AN ITALIAN NEWBORN POPULATION.

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The high incidence of HIV infection in females of reproductive age in Italy is responsible for a high number of pediatric AIDS cases compared to other European Countries. Therefore it is important to assess the prevalence of HIV infection among parturients in order to estimate the further incidence of pediatrics AIDS in the population. Between June 1988 and November 1989, 67,337 blood samples collected from consecutive newborns for routine metabolic screenings in 98 hospitals from different Italian regions were examined for HIV-Ab. Blood saturated disks were punched out from the collection paper, without identification, eluted in citrate buffer and screened for HIV-Ab in ELISA: the positive results were confirmed in a Western Blot. Among the 67,337 blood samples examined 82 (0.12%) - (95% Confidence Limits 0.097--0.151) were positive for HIV-Ab. The distribution pattern of positive samples among the different regions correlated with the local pediatric cumulative incidence rate, with higher values in Lombardia and Lazio regions. Anonymous serosurveys for HIV-Ab in newborns provide useful data for the estimation of HIV prevalence in the population and for monitoring the trend of the infection in females of reproductive age.