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CONTRIBUTION TO THE STUDY OF SCID PATHOGENESIS DUE TO ADA DEFICIENCY. EFFECT OF 2' DEOXYADENOSINE COMBINED WITH 2' DEOXYCOFORMYCINE ON NEONATAL LYMPHOCYTE PROLIFERATION.

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It has been demonstrated that an accumulation of deoxyadenosine (dAd) in adenosine deaminase (ADA) deficient individuals proves to be markedly toxic for lymphoid cells. The immunological function of such children is usually normal at birth, apparently due to the transplacental removal of toxic metabolites during gestation. We studied the possibility that this immunological normality might also be due to a different lymphocyte physiology in neonates. To do so, we cultured mononucleated cells from cord blood of 33 term-born neonates and added to the cell suspensions different concentrations of dAd after inhibiting ADA activity with deoxycoformycin. We observed that in this artificial situation of ADA deficiency dAd significantly inhibits the proliferation of cord blood lymphocytes, both regarding the response to PHA ($19.99 \pm 5.81\%$ of the response in control cultures) and to ConA ($15.73 \pm 3.17\%$). However, the inhibitory effect was significantly smaller ($p < 0.01$) than that observed in controls ($7.79 \pm 2.91\%$ with PHA and $8.10 \pm 0.78\%$ with ConA). The different response may be due to the different proportion of cord blood cell populations but also to intrinsic differences which might contribute to the delay in the clinical expression of this enzymatic deficiency.

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ACTION OF 2' DEOXYCOFORMYCIN ON MITOGEN-INDUCED LYMPHOPROLIFERATION IN THE NEONATAL PERIOD.

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The effect of 2' deoxycoformycin (dCF), a potent inhibitor of adenosine deaminase (ADA), on the proliferation of mononuclear cells from cord blood and from healthy controls as a response to the two mitogenic agents PHA and ConA was studied. The addition of dCF simultaneously with the mitogen did not modify cell proliferation either in neonates or in controls. Added 20 min before the mitogens, dCF induced in adult lymphocytes a significant inhibition in the response to PHA ($68.11 \pm 10.40\%$ of response in control cultures) and to ConA ($58.78 \pm 26.23\%$). By contrast, in the neonatal period it induced a stimulatory effect on this response, both when PHA ($117.64 \pm 26.48\%$ of basal response) and ConA ($108.18 \pm 21.72\%$) were employed. The possibility is discussed that this different function in lymphocyte behaviour in the newborn might contribute to the immunological normality which children affected with ADA deficiency may exhibit at this age and also to the delay in the onset of clinical manifestations of immunodeficiency due to the defect in this enzyme.

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RELEVANCE OF IN VITRO METHODS FOR THE DIAGNOSIS OF ALLERGY TO ALTERNARIA TENUIIS AND CLADOSPORIUM HERBARUM

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In 22 patients suffering from allergic asthma caused by *Alternaria tenuis* and/or *Cladosporium herbarum* as proven by bronchial provocation tests the following diagnostic procedures were performed: skin prick test determination of specific IgE antibodies by RAST (comparing old type RAST discs with new more sensitive mould discs) and the detection of specific IgE and IgG antibodies using an immunoblot technique. Finally, mould induced histamine release was analysed. In only 34% skin test was found to be positive. Old type RAST discs detected specific IgE antibodies in 70%, whereas the new improved discs were able to show IgE antibodies in 88%. By means of immunoblotting specific IgE and IgG were demonstrated in most of the patients. The best correlation to the state of allergy, however, was achieved by mould induced histamine release, which was positive in 94%. The data show that the use of new improved mould discs for IgE RAST, but also of mould induced histamine release and immunoblotting strongly increase the possibilities for detection of allergy against *Alternaria* and *Cladosporium*.

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SPECIFIC IMMUNOTHERAPY (SI) IN THE TREATMENT OF "MINOR" ALLERGIC RESPIRATORY DISEASE (MARD) (wheezy bronchitis and/or chronic cough)

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We studied 224 children (ch) with high suspicion of MARD, aged 1 to 13 ($x=4.6$ yr) with more than 3 acute episodes during more than 6 months. We found an allergy family history in 71.9% and a high level of total IgE in 92%. We found positive results to house dust mites in 43.7% with prick test and in 51.8% with RAST. Both PT and/or RAST gave positive results in 68.7%. The follow-up study was conducted over 1 to 4 yr ($x=2$ yr); 37 dropped out. We divided 150 proven atopic ch into 3 groups. Group A (69 ch) received a mast cell stabilizer drug (MCS), group B (45 ch) SI with D.Pteron allergen plus MCS, group C (36 ch) served as a control group. A marked improvement was seen in group B, whereas in group C only 1 of 6 ch showed any improvement. Group A showed intermediate results. During the study 50% of group C developed overt asthma against 30% in group A and only 4% ($p < 0.01$) in group B. The results suggest that SI might have a preventive role in children with atopic MARD.

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MID-TERM OUTCOME IN 46 SURVIVORS WITH MAJOR PERI-INTRAVENTRICULAR HEMORRHAGE (PV-IVH).

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From Feb.1981 to Dec.1985 (4 years 10 months), 2400 neonates were admitted to Port-Royal NICU, 1/3 being <1500 g. Ultrasound scanning detected 392 PV-IVH, of which 130 were major H. (from unilateral grade III to bilateral grade IV H.). Survival rate was 65% in unilateral III H., 33% in bilateral III H., 32% in unilateral IV H., 1/13 bilateral IV H. The 46 survivors with major PV-IVH were followed for over 1 year in 39 cases (3 were lost at 3 mths and 4 are under 1 year as of writing), up to 5 years. Among 14 unilateral III H., 2 infants were lost, 1 is <1 y., 9 were normal, 2 had minor anomalies. Among 17 bilateral III H., 1 is <1 y., 6 were strictly normal, 4 had minor anomalies, 2 had a moderate impairment, 4 had severe handicaps. Among 14 unilateral IV H., 1 infant was lost, 2 are <1 y., 6 were strictly normal, 1 had minor anomalies, 3 had a moderate impairment, 1 had a severe handicap. The only survivor with bilateral IV H. was severely handicapped by 4 years of age. The overall summary in 39 cases with accurate data over 1 year totals 21 strictly normal infants, 7 with minor anomalies, 5 with moderate and 6 with severe sequelae. The results will be up-dated and completed at Congress' date.