Abstracts

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SERUM THYMIC FACTOR (SF) IN PRIMARY IMMUNODEFICIENCY DISEASES

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Direct measurements of SF was performed by a cAMP assay using mouse thymocytes (1) in 15 patients with several forms of infantile immunodeficiency diseases. In addition we investigated, among patients' lymphocytes, the presence of "target cells" for SF, i.e. lymphocytes which could be stimulated by SF to increase intracellular cAMP, which is a property of precursors of mature T cells. Results were the following:

disease	SF	target cells	disease	SF	target cells
SCID	+	-	T cell defect		
SCID	+	-	with PNP def.	+	-
SCID	+	n.d.	thymic displ.	-	n.d.
SCID	-	-	unsp. T cell		
CID	-	n.d.	defect	+	n.d.
CID	-	n.d.	Wiskott-Aldrich	-	n.d.
di George	-	+	Cartilage-hair		
incompl.			hypopl.	±	+
di George	±	-	Ataxia-tel.	-	+
			Ataxia-tel.	-	n.d.

The detection of SF activity and of "target cells" for SF provides an additional aid for diagnosis of primary immunodeficiencies and a way to evaluate thymic function also in view of treatment with transplantation(s) and/or thymic hormone(s).

1) Astaldi, A. et al., Nature 260, 713-715 (1976).

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GRANULOPOIESIS IN THE HUMAN EMBRYONIC LIVER: DETECTION OF GRANULOCYTIC PROGENITOR CELLS BY IN VITRO METHODS

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Since hematopoiesis in the human embryonic liver is almost exclusively erythropoietic, without any appreciable evidence for granulopoiesis, it was attempted to demonstrate granulocytic progenitor cells in this organ, by virtue of their capacity to form in vitro granulocytic-Monocytic colonies in soft agar cultures. 31 human embryos were obtained by interruptions of pregnancies at 6–12 weeks of gestation. Liver cell suspensions were seeded in soft agar over feeder layers composed of normal human leukocytes. Colonies were counted and examined by morphologic, cytochemical and electron microscopic methods following 7 days of incubation. A clonning yield of 60 ± 28 colonies/2x10⁵ cells seeded was obtained in the stimulated, and none in the non-stimulated cultures, comparable to the findings in cultures of adult human marrows. No significant variations in the growth rates of the colonies at different gestational ages were observed. A layer of mononuclear cells was obtained by fractionation of the liver cell suspension over Ficoll-hypaque gradient, which yielded a 5 to 10 fold increment in the clonning efficiency. While only scarce granulocytic and monocytic elemeats were detected in the liver cell suspension prior to culture, the colonies obtained were almost exclusively granulocytic with normal maturation as was evident by cytochemical and electron microscopic parameters. These studies have shown that human embryonic liver at 6–12 weeks of gestation contains an abundancy of granulocytic progenitor cells comparable to adult marrow in their quantity, density, dependence on colony stimulating factor and capacity to differentiate into their mature progeny. The possible potentiality of human embryonic liver for human hematopoietic cell transplantation is suggested by these results.

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SPLENECTOMY and INFECTION : A FOLLOW-UP OF 34 CASES

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In a group of 34 children(ages 6 mos.–20 yrs.)who were splenectomized due to trauma(15 cases) and non-trauma(19 cases of ITP, beta thalassemia, hereditary spherocytosis, portal hypertension, hypersplenism), the absolute lymphocyte counts, E rosettes, skin tests(PHA, candida, SKSD, PPD), isohemagglutinins and serum IgG levels were not different than those of 34 age matched controls ($P > 0.05$). However serum IgA and IgM levels were low ($P < 0.05$). Fifteen patients(44%) developed infections(bronchopneumonia, hepatitis, wound abscess, sepsis) following splenectomy. A defective opsonization for pneumococcus type II-L was found in 8. Six of these had various infections. Pre and post splenectomy immunoglobulins determined in 11 cases showed low IgA and IgM levels in 8 and 9 patients respectively. Our study also confirms the high incidence of infection regardless of the cause for splenectomy since 33.3% of the patients in traumatic and 52.6% of the patients in non-traumatic group had infection.

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REDUCED CELLULAR IMMUNITY WITH NORMAL IMMUNOGLOBULINS AND A DEFECTIVE ANTIBODY PRODUCTION IN TWINS

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The primary immunodeficiency diseases represent themselves with a broad spectrum but sometimes reaching a diagnosis is difficult because of mild symptoms and normal tests. An example of this was a set of twins in whom various immunological tests (blastic transformation with PHA, skin tests, serum immunoglobulins, etc) were within normal limits. They had recurrent respiratory infections and gastroenteritis necessitating 9 hospitalizations over a period of 17 months. Their 11 siblings died before 9 months of age with a history of similar infections. The autopsy showed a thymic dysplasia and depletion of the lymphocytes in thymus dependent areas and the presence of plasma cells in the peripheral lymphoid organs of one twin who died of sepsis. The other twin did not reject a skin graft and showed depletion of lymphocytes in the paracortical areas of an antigen stimulated inguinal lymph node. His red cell ADA and nucleotide phosphorylase levels, EA, EAC rosettes, surface immunoglobulins were within normal range. His antibody response to KLH and diphteria-tetanus toxoids was poor. His primary and secondary antibody responses were both decreased to bacteriophage ØX,174 although he cleared the antigen within 6 days and had a memory amplification. Fifty three per cent of the antibody was of the IgG class. Thymosin(in vitro) caused a significant increase (48% of the initial value) in E rosettes. This was not observed in the healthy controls.