733 ANTIBODY RESPONSE TO INTRAVENOUS IMMUNIZATION FOLLOWING SPLENIC TISSUE AUTOTRANSPLANTATION IN SPRAGUE-DAWLEY RATS. Allen D. Schwartz, Mahboubeh Dadash-Zadeh, I. Richard Goldstein, Susan Luck, and James J. Conway. University of Maryland School of Medicine, Department of Pediatrics, Baltimore, and Children's Memorial Hospital, Departments of Pediatrics, Surgery, and Radiology, Chicago.

Autotransplanted splenic tissue has been shown to regenerate into implants microscopically indistinguishable from normal spleen in a variety of experimental animals. Little is known, however, about the immunologic function of such implants. The spleen is essential for the formation of antibody in response to small doses of intravenously administered particulate antigen. Response to intravenous challenge with sheep erythrocytes was determined in 12 Sprague-Dawley rats 5 months following autotransplantation of splenic tissue into subcutaneous tissue, peritoneal cavity, or a surgically-created omental pouch. There was a marked rise in heterophile antibody titer following intravenous challenge in 10 control animals and no rise in 9 of 10 asplenic animals. Heterophile antibody titers increased in all of the animals with transplants. In addition, autotransplanted splenic tissue was able to take up intravenously injected ^{99m}Tc sulfur colloid. Thus autotransplanted splenic tissue is capable of clearing intravenously administered particulate material from the circulation and is able to have an immunologic response to intravenous antigenic challenge similar to that of the normal, intact spleen.

734 REGENERATION OF T-LYMPHOCYTE RECEPTOR FOR SHEEP ERYTHROCYTES. Michael R. Sharpe, Nicholas DiVito, and Raymond D. A. Peterson. University of South Alabama College of Medicine, Department of Pediatrics, Mobile.

The formation of erythrocyte (E)-rosettes by human T-lymphocytes with sheep erythrocytes (SRBC) is the most prevalent method by which T-lymphocytes are quantitated and distinguished from B-lymphocytes. The factors responsible for the binding of SRBC to T-lymphocytes are unknown. The present study was done in order to clarify the physiochemical basis of this cellular interaction, specifically as regards factors that influence the regeneration of SRBC receptors on T-lymphocytes after their removal by trypsin treatment. After exposure of lymphocytes to 0.1% trypsin for 5 minutes, the percentage of rosette forming cells was decreased to less than 10%. After 18 hours incubation at 37° in HBSS, complete regeneration has occurred so that the percentage of rosette forming lymphocytes is not significantly less than values for non-trypsinized cells. Regeneration is inhibited by incubation of lymphocytes with puromycin 20 µg/ml and cyclohexamide 10 µg/ml to levels that are 7% and 33% respectively of values for cells that have regenerated their receptors. The T-lymphocyte receptor for SRBC appears to be a protein structure that is rapidly resynthesized in the absence of serum factors. (Supported by Kiwanis Cancer Research Grant)

735 IN VITRO AND IN VIVO DEFINITION OF A NEW VARIANT OF SEVERE COMBINED IMMUNODEFICIENCY DISEASE (SCID) Abraham Shore, Hans-Michael Dosch, Johannes Huber and Erwin W. Gelfand, The Hospital for Sick Children, Departments of Immunology and Pathology, Toronto, Canada

SCID is a heterogeneous disease caused by at least two known primary defects; specific enzyme deficiency or a defect in thymic epithelial cell maturation. A 12 month old male presented with a 7 month history of recurrent fevers, pyogenic infections, and documented Pneumocystis carinii pneumonia. Examination revealed hyperplastic lymphoid tissue. Laboratory data included normal red cell enzymes, normal lymphocyte numbers, profound hypogammaglobulinemia (except for an IgM of 22-47 mg%), no antibody response, but normal numbers of B-cells. 9-15% of his peripheral blood lymphocytes formed E-rosettes; both his PHA and MLR responses were markedly reduced (10% of normal). He did not respond to several skin test antigens or DNCB, and he failed to reject an allogeneic skin graft. Lymph node biopsy demonstrated normal follicle formation, poor germinal center formation, and depletion of cells in the paracortex. Thymus biopsy revealed a large thymus with predominantly E-rosetting cells, PAS-positive epithelial cells, but no Hassall's bodies. Unlike other SCID patients, bone marrow precursor cells were responsive normally to human thymus conditioned medium, suggesting the initial stage(s) of T-cell maturation were achieved in this patient. On the basis of the thymus biopsy and bone marrow precursor cell response, we believe this case of SCID represents a failure of terminal differentiation of intrathymic T-lymphocytes.
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736 RECOVERY OF MACROPHAGE C3 RECEPTOR FUNCTION FOLLOWING SELECTIVE ENZYMATIC DESTRUCTION.

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To examine macrophage receptor metabolism, rabbit pulmonary macrophages were exposed to trypsin (1mg/10^7 cells) for five minutes. This reduced C3-activated ingestion by 70%, but did not affect nonspecific or IgG-mediated ingestion. Trypsinized cells replaced their C3 receptors completely when incubated for four hours in media containing fresh or heated serum or 4% bovine albumin (BSA) at 37°, but not in buffered salt solutions with or without glucose and amino acids, gelatin solutions, or in any medium at 0°. Cycloheximide did not affect phagocytosis at a concentration of 2 ug/ml, but abolished ^{3}H-leucine incorporation into protein, and prevented recovery of the C3 receptor of trypsinized cells. Rates of protein synthesis by macrophages were equivalent in the presence or absence of serum. We conclude that there are two independent requirements for receptor recovery: (1) endogenous protein synthesis, and (2) a heat-stable exogenous component present in serum and high concentrations of BSA.

737 LYMPHOCYTIC INTRACYTOPLASMIC INCLUSIONS IN INFANTS WITH A VARIANT OF SEVERE COMBINED IMMUNODEFICIENCY (SCID). O.F. Sieber, Jr*, J.F. Jones*, C.M. Payne* and V.A. Fulginiti, Departments of Pediatrics and Pathology, University of Arizona, Tucson, Arizona.

SCID as a genetic disease is inherited either as an autosomal or a sex-linked recessive with an incidence of 1:2,000,000 or 1:10,000, respectively. Over a six year period, we have identified 8 infants from a single ethnic group with a SCID rate greater than 1:3500. Three infants were born during a recent 3 month period, only one with a family history of SCID. In addition to a high group frequency and unusual clustering of cases, this SCID syndrome variant included a failure to clear ØX174 from the blood, an absence of a common HLA type or a history of consanguinity. By electron microscopy, a large percentage of peripheral blood lymphocytes contained cytoplasmic inclusions classified as parallel tubular arrays (PTAs), occurring in both E and EAC rosetting cells. The average number of inclusions was 15 times greater in these infants (range 13-53%) than in lymphocytes from age matched control infants (range 0-7%). Similar inclusions in as high a number have been described in Hodgkin's disease, chronic lymphocytic leukemia and Epstein-Barr virus (EBV) infections, diseases which also have occurred in clusters. No evidence for EBV was determined serologically in any infant or in 2 of 3 mothers. In one infant successfully transplanted, PTAs have decreased in number. Explanations possible for the PTAs and the SCID variant are: 1) PTAs are abnormal metabolic organelles; or 2) are viral inclusions associated with SCID; or 3) are specific acquired virus.

738 DYSKERATOSIS CONGENITA WITH PANCYTOPENIA (DC-P): A MONOCYTE DEFECT? Clark M. Smith II, Robert D. Nelson†, Vance D. Fiegel*, Norma K.C. Ramsay, Mark E. Nesbit and William Krivit. University of Minnesota School of Medicine Departments of Pediatrics, Laboratory Medicine†, and Surgery*, Minneapolis.

Dyskeratosis Congenita (DC) is a rare hereditary syndrome of ectodermal dystrophy (ungual dystrophy, poikiloderma atrophicans, leukoplakia) in which ½ of the reported cases have had pancytopenia (DC-P). These patients have an excessive incidence of infection presumably related to loss of normal mucous membrane barrier, pancytopenia and steroid therapy. We will report two more unrelated typical DC-P phenotypes with special attention to leukocyte function in one. Although previously not reported, both patients had bone marrow responses to oxymetholone within 3 months (erythroid>myeloid >megakaryocyte). Pt 1 had a low normal IgM of 51mg%: he expired from sepsis with an absolute neutrophil count of 3,000. Pt 2 has a low IgM of 27mg%: he has had less ungual pain on zinc and remains free of clinical infections. His monocyte function studies before oxymetholone, on zinc, demonstrated depressed spontaneous migration (50% of control, twice), no chemotactic response to zymosan activated serum (twice), and depressed chemiluminescence (65% of control, once). Response to MIF and production of colony stimulating factor were normal. Neutrophil function studies, lymphocyte mitogen response, and mixed lymphocyte culture stimulation were normal. Zinc therapy would not seem to be an explanation for these in vitro findings given the rapid egress of zinc from monocytes and the experimental conditions used. A primary monocyte defect may be present in DC-P.