

902 MONOCYTE SUPPRESSION OF IMMUNOGLOBULIN SECRETION IN A 9-YEAR-OLD BOY WITH SCID. Lewis A. Brown, E. Clinton Lawrence and William T. Shearer, Depts. of Pediatrics and Medicine, Baylor College of Medicine, Houston, TX 77030

We report the evaluation of *in vitro* immunoregulation in a 9-year-old untreated boy with SCID. Fico11-Hypaque-isolated peripheral blood mononuclear cells (MNL) from the patient failed to respond to pokeweed mitogen (PWM) with the normal increment in immunoglobulin-secreting cells (Ig-SC), as measured by a reverse hemolytic plaque assay. Removal of phagocytic cells or the addition of unrelated irradiated helper cells resulted in enhanced, but still suboptimal response to PWM, suggesting some intrinsic defect in B-cell function. Co-culture of patient with normal MNL resulted in marked (88%) suppression of PWM-induced Ig-SC. Suppressor activity was unaffected by prior irradiation of patient MNL, but was substantially reversed (32% net enhancement) by removal of his phagocytic cells; whereas the combination of the two procedures further reversed suppression (52% net enhancement). Because the patient was lymphopenic, his MNL were relatively enriched for monocytes (range=40-80%). To determine whether the suppressor cell activity was due to functional or numerical excess of patient monocytes, co-cultures were performed using varying ratios of patient to normal MNL. Suppression was most marked in cultures containing >40% monocytes, suggesting that a numerical excess of patient monocytes could account for the observed suppression. These data suggest both an intrinsic defect in B-cell function, and a relative numerical excess of monocytes which could further inhibit Ig-secretion by B-cells.

903 IMMUNE COMPLEXES IN DELAYED ONSET FOOD ALLERGY. Terrance T. Chang, Devron Char, and Oscar L. Frick, University of California, San Francisco, Department of Pediatrics, San Francisco, CA.

Circulating immune complexes (CIC) have been implicated in the pathogenesis of adverse reactions to foods. Two immune complex assays, Raji cell and Clq-binding were used to test 25 sera from patients with delayed onset food allergy (DOFA). Raji cells bind CIC via C3, C3b, Clq and Fc-IgG surface receptors. Greater than 40 µg/dl is considered a positive test. A solid phase Clq assay with ¹²⁵I-Staphylococcus A Protein was also used to measure CIC. The percentage of counts bound by patient's serum was compared to a human cord sera pool; a positive test was >1.5X.

Raji cell assay was positive in 18/25 (72%) of patients with DOFA; 9 of these (36% of total) had >100 µg/dl - a range commonly observed in patients with SLE. Clq-binding was positive in 13/25 (52%). In 18/25, there were coincident results in both Raji and Clq-binding; in another 4, there was concordance over a wider range, and in 3, there was discordance. In 14 patients with selective IgA deficiency, 8/14 had positive Raji and 6 of these had positive cow's milk precipitins. Leukocyte inhibition factor (LIF) was positive for one or more foods in 10/12 DOFA patients tested. In the 25 DOFA patients, immediate skin tests with foods were positive in 2, and IgE-RAST with foods was positive in 7. Twenty age-matched non-allergic children had negative Raji and Clq-tests. These results suggest that IgG immune complexes may be involved in the pathogenesis of DOFA to account for symptoms which may frequently resemble those of serum sickness.

904 DOUBLE LIGHT CHAIN PRODUCTION BY LEUKEMIC CELLS OF COMMON CLONAL ORIGIN. Young J. Choi and Martha Wong, Albert Einstein College of Medicine, The Bronx-Lebanon Hospital Center, Departments of Pathology and Pediatrics, Bronx, New York. (Sponsored by M. Cohen).

Generally lympho-plasmacytic malignancies are of monoclonal origin in that neoplastic B cells produce immunoglobulin (Ig) composed of a single class of heavy and light chains. Thus the presence of more than one monotypic Ig on the surface and in the cytoplasm of the neoplastic cells of a patient constitutes an uncommon event. We are presenting an 11 year old boy with acute lymphoblastic leukemia of the rare B-cell type in which the same neoplastic cells produced both κ and λ light chains without any heavy chains as demonstrated by double immunofluorescence. There was evidence of secretion of both κ and λ light chains in cultures of the leukemic cells. Secretion of both light chains into the blood was also demonstrated at postmortem examination. Although the production of more than a single class of Ig has been known to occur in rare cases of multiple myeloma and Waldenstrom's macroglobulinemia, such an event has not been reported in patients with leukemia or lymphoma.

905 CHILDHOOD MIXED CONNECTIVE TISSUE DISEASE. David S. Chudwin, Diane W. Wara, Morton J. Cowan, Arthur J. Ammann, University of California, Department of Pediatrics, San Francisco.

We reviewed the clinical and laboratory features of 6 children with MCTD (4 girls, 2 boys). Initial symptoms (mean age of onset 9.2 years) were arthritis (2), chronic fevers (2), and urticaria and Raynaud's phenomenon (2). Mean age at diagnosis was 14.2 years. Clinical features included:

6/6 Raynaud's phenomenon	6/6 Chronic headaches
5/6 Arthritis	6/6 Depression
4/6 Chronic fevers	4/6 Sjogren components
4/6 Chronic dermatitis	4/6 Dysphagia
3/6 Sclerodermatous skin	2/6 Pleuritis, pericarditis
2/6 Urticaria	2/6 Hepatosplenomegaly
3/6 Alopecia	2/6 Lymphadenopathy

Laboratory findings were as follows:

6/6 Speckled ANA	2/4 Renal biopsy abnormal
6/6 Ribonucleoprotein titer $\geq 1:102,400$	4/6 Esophogram abnormal
3/6 Hematuria	4/6 Pulmonary tests abnormal
3/6 Proteinuria	1/6 Hypocomplementemia
3/6 + creatinine clearance	3/6 Hypergammaglobulinemia
	2/6 Thrombocytopenia

There has been no mortality, but 3/6 are significantly disabled (mean followup of 3.4 years) despite treatment with salicylates, prednisone and in 3 patients chlorambucil. Our experience with MCTD in children reflects greater chronic morbidity than reported in adults.

906 IMMATURE PHENOTYPE OF IGA B CELLS FROM IGA DEFICIENT PATIENTS. Mary Ellen Conley and Max D. Cooper, University of Alabama Medical School in Birmingham, Cellular Immunobiology Unit, Department of Pediatrics and Microbiology, Birmingham, Alabama 35294

IgA B cells have been detected in the peripheral circulation of most patients with IgA deficiency. It has recently been shown that the surface isotypes expressed by human IgA B cells vary as a function of their stage of maturation. To determine the stage at which IgA B cell differentiation is blocked in patients with IgA deficiency, peripheral blood lymphocytes from 11 patients with serum levels of IgA < 2 mg/dl were stained for surface immunoglobulins by immunofluorescent techniques. Cells expressing surface IgA were detected in all patient samples but occurred at a significantly lower frequency than in samples from normal controls (0.09% vs. 1.4% of the total lymphocytes). In 10 of the 11 patients 87% of the IgA B cells co-expressed IgM (range 70-100) and 81% expressed IgD (range 65-94). This staining pattern was similar to that seen in the normal newborn in which 95% of the IgA B cells expressed IgM and IgD. In contrast only 5% of the IgA B cells from normal adults co-expressed IgM and IgD. The remaining IgA deficient patient had only 0.05% IgA B cells but these cells demonstrated a typical adult phenotype. 3% of the IgA B cells bore IgM and none bore IgD. These results indicate that in most patients with IgA deficiency IgA B cells differentiation is blocked at an early stage. (Supported by NIH Grants CA16673 and 5MOR32 and National Foundation Grant 1-608).

907 DILANTIN-INDUCED SPONTANEOUS SUPPRESSOR (T) CELL ACTIVITY AND HYPOGAMMAGLOBULINEMIA. Dosch, H-M., Jason, J. and Gelfand, E.W., Division of Immunology, Research Institute, Hospital for Sick Children, Toronto, Ontario.

Dilantin therapy has been associated with immunoglobulin deficiency. An 11 year old male, treated for 8 months with dilantin for post-traumatic seizures, presented with an acute onset of fever, rash and diffuse lymphadenopathy. Serum IgA, IgM and IgE were virtually absent and IgG levels were very low (200 mg.%); circulating B-cells were markedly reduced with less than 0.2% of the mononuclear cells stained for sIgM/sIgD; T-cell numbers were normal. Using an antigen-specific hemolytic plaque assay, it was found that fresh, patient T-cells were able to suppress immunoglobulin secretion in normal as well as autologous B lymphocytes, sensitized *in vitro*. This spontaneous suppressor activity, previously described in some patients with X-linked agammaglobulinemia, was inhibited by lithium, a putative blocker of cyclic AMP generation. (J. Immunol. 121: 2097, 1978). Two weeks following cessation of dilantin therapy, clinical signs of illness disappeared, suppressor cells were no longer detectable and B lymphocytes began to re-appear. Normal numbers of circulating B lymphocytes were restored by 4 weeks and serum IgG, IgA, IgM and IgE levels were normalized by 8 weeks. These data suggest that in susceptible individuals, dilantin therapy may result in hypogammaglobulinemia. The coincidence of drug-induced antibody deficiency and the emergence of spontaneous suppressor cells supports the concept that abnormal suppressor T cells may play a role in certain antibody deficiency syndromes.