**739** DEFECTIVE IN VITRO LYMPHOCYTE RESPONSE TO PSEUDOMONAS AERUGINOSA IN SEVERELY ILL CYSTIC FIBROSIS PATIENTS by Ricardo U. Sorensen, Robert C. Stern and Stephen H. Polmar, Case Western Reserve University, Department of Pediatrics, Cleveland, Ohio.

Pulmonary infection with Pseudomonas aeruginosa (PA) eventually occurs in virtually every patient with cystic fibrosis. Progression of infection and, ultimately, death occurs despite circulating specific antibody. For these reasons, studies of cell mediated immune responses to PA were undertaken in patients with cystic fibrosis. In vitro lymphocyte proliferative responses to PA, Hemophilus influenzae, Streptococcus hemolyticus and Staphylococcus aureus antigen were tested by $^{3}H$ thymidine incorporation. Twenty-nine patients with chronic PA infection including 16 in fair or good condition (Shwachman Case History Score of 15-25 points) and 13 in poor condition (Score of 1-14) were tested. In addition, 6 patients who had never had PA recovered from sputum cultures and 13 normal persons were also tested.

The mean response to three different PA strains was 753 counts per minute (cpm) in the "poor" group compared to 3234 cpm in the "good" group ($p< 0.0005$) and 2572 in the normal groups ($p<0.005$). This difference was not present in the responses to PHA and Con-A or to the other bacterial antigens. Two terminally ill patients gave lower responses to four of their own PA strains than did the other patients. In addition, two sibling pairs in which one child was severely ill and infected with PA while the other child had not yet shown PA on sputum cultures were studied. In both cases, the uninfected child had normal responses, wherease the severely ill sibling had virtually no response against his own PA.

**740** DIFFERENCES IN ENERGY METABOLISM OF MONONUCLEAR CELL SUB-POPULATIONS. S. Storch, W. Klein, M. Das, and S.A. Feig, UCLA Sch. of Med., Dept. of Peds., L.A.

Impairment of cell mediated immune (CMI) function has been correlated with abnormalities of mononuclear leukocyte energy metabolism. In order to improve the understanding of the metabolic basis of immune function, we have compared glycolytic metabolism of two sub-populations of normal mononuclear cells, T- and non-T cells. Mononuclear cells were isolated from blood by density-gradient centrifugation. Enrichment of T-cells was accomplished by nylon column separation; non-T cells were harvested from the interface of a density gradient after rosetting with sheep erythrocytes. Glucose utilization (GU) by non-T cells ($206\pm33$ $\mu moles/10^{10}$ cells/hr) was much greater than that by T-cells ($23\pm15$) ($p<0.001$). The addition of PHA to T-cell incubation caused an immediate four-fold stimulation of GU while non-T cells were totally unaffected ($92\pm22$ vs $196\pm20$). Lactate production (LP) by enriched T-cells ($34\pm19$) was much less than LP by non-T cells ($351\pm35$) ($p<0.001$). The addition of PHA to the incubation stimulated LP by T-cells ($104\pm35$) but did not affect LP by non-T cells ($316\pm36$). The ATP content of T-cells was identical to that in non-T cells ($6.4\pm2.0$) and remained stable during the course of the incubations. The metabolic activity of sub-populations of mononuclear cells is consistent with observations of metabolism in unseparated mononuclear cells. These studies document marked differences in basal and stimulated metabolic activity of mononuclear cell sub-populations, and suggest new approaches toward the understanding of the metabolic basis of immune dysfunction.

**741** OXIDATIVE METABOLISM IN HUMAN CORD BLOOD NEUTROPHILS. Ronald G. Strauss and M. J. Seifert, University of Iowa Hospital, Department of Pediatrics, Iowa City, and St. Jude Children's Research Hospital, Memphis.

Oxidative metabolism (OxM) was assessed in cord blood neutrophils (CBN) in an attempt to explain the susceptibility of infants to infections. A variety of methods were employed in prior reports, and results have been contradictory. Isolated neutrophils were used in this study. Hexose monophosphate shunt (HMS) activity was determined by glucose-1-$^{14}C$ oxidation. The kinetics of OxM were measured as the rate of light emission by chemiluminescence (CL), an assay related to superoxide and singlet oxygen formation. HMS activity in both resting and phagocytic cells was greater ($p < 0.05$) in 17 CBN than in cells from 9 simultaneously studied adult controls, however, the absolute increase from resting to phagocytic values was significantly less in CBN. Mean values for peak CL during phagocytosis of zymosan from 14 CBN and 9 controls were similar (18.0 vs 20.7 CPM x $10^4$, $p > 0.05$), but the kinetics of CL were quite different. During the first 8 min. of phagocytosis CL (light emission) increased sharply in both groups. At later times CL was sustained by control cells, but waned in CBN and within 15 min. was significantly less ($p < 0.05$). Thus, CBN initiated post-phagocytic OxM and reached normal peak values of HMS activity and CL. However, CBN were unable to maintain the increased rate of OxM after phagocytosis when compared to adult controls. This difference may indicate a functional defect.

**742** RICE ALLERGY WITH CNS REACTIONS AND SERUM COMPLEMENT DEPRESSION. Robert C. Strunk, Jacob L. Pinnas, T. Jacob John, Ronald C. Hansen, Julie L. Blasovich. Dept. of Ped. & Med., Univ. of AZ, Tucson, AZ. (Intr. by James J. Corrigan, Jr.)

Rice allergy has been reported, but not adequately documented. An infant had flushing, vomiting and diarrhea after his first and 6 subsequent rice feedings from 2 to 15 months of age. In addition, ingestion of rice or soy during his first 6 months induced lethargy, unresponsiveness and EEG abnormalities. RAST and P-K testing documented IgE antibody to rice, but not to soy. Rice fluorescent antibody spot tests for anti-rice antibody were positive for IgE, but negative for IgG, A & M. Precipitation and passive hemagglutination antibody studies were negative for rice and soy. Early in the course serum complement (C) was depressed both before and after rice challenge. CH50, C3, C4 and Factor B levels remained low for 2 months and did not rise to normal until rice and soy were eliminated from diet at 6 months of age. Rice ingestion at 13 months of age was followed by flushing, vomiting and diarrhea and was associated with increases of serum histamine of 140% at one hour and 70% at eight hours after challenge. There was no associated CNS reaction or C depression with this challenge. The CNS reactions and C depression prior to six months of age may have been due to rice allergy combined with gut and/or CNS immaturity. Rice allergy was demonstrated by specific IgE and GI reactions to rice even after C returned to normal. This case documents true rice allergy in which C depression and CNS reaction accompanied the GI reaction.

**743** ESTABLISHMENT AND CHARACTERIZATION OF LYMPHOBLASTOID CELL LINES (LCL) DERIVED FROM PATIENTS WITH IMMUNE DEFICIENCY SYNDROMES (IDS). John L. Sullivan, Hans D. Ochs, Carol L. Dunsmoor, and Ralph J. Wedgwood. Department of Pediatrics, University of Washington, Seattle.

Using Epstein-Barr virus (EBV), we have established LCL from 15 patients with IDS, including patients with ataxia telangiectasia (3), Wiskott-Aldrich syndrome (1), severe combined immune deficiency (1), common variable immune deficiency (5), X-linked immune deficiency with increased IgM (1), immune deficiency with normal immunoglobulins (1), C4 deficiency (1), Job's syndrome (1). LCL could not be established from 4 patients with infantile X-linked agammaglobulinemia despite multiple attempts. All LCL have surface markers and functions characteristic of normal B-lymphocytes, including C3 receptors and surface immunoglobulins, and were potent stimulators in the mixed lymphocyte reaction. None of the LCL expressed T-cell characteristics. The inability to establish LCL from X-linked agammaglobulinemic patients might reflect absence of EBV receptors as well as absent B-lymphocytes. Most of the LCL appear to be polyclonal. Individual lines expressed more than one class of surface immunoglobulin. Indeed, LCL were established which expressed immunoglobulin classes markedly deficient or absent in the serum of the patients from which they were derived, suggesting the defect resulted from suppressor activity rather than defective synthesis. Further immunologic and metabolic studies of these lines may help to elucidate pathologic mechanisms responsible for primary IDS.

**744** CELL MEDIATED IMMUNE RESPONSE TO LIVER SPECIFIC ANTIGENS FOLLOWING HEPATITIS B VIRUS INFECTION IN MAN. M.L. Tiku, K.R. Beutner, K. Tiku, P.L. Ogra. SUNY at Buffalo, New York.

The in-vitro correlates of cell mediated immunity (CMI) against liver specific antigens (LSA) and hepatitis B surface antigen ($HB_sAg$) was studied in groups of subjects with acute or chronic hepatitis B virus (HBV) infection and in a population of uninfected healthy controls. The techniques of in-vitro lymphoproliferative responses (LTF) to $HB_sAg$, LSA, phytohemagglutinin (PHA) were employed in these studies. The LSA consisted of liver tissue extracts prepared from HBV seronegative autopsy materials. No LTF response to LSA and $HB_sAg$ was observed in control population. All patients with acute HBV infection manifested significant LTF response to $HB_sAg$ as well as to LSA. Peak responses were observed at the height of clinical disease. 50% of patients with chronic $HB_sAg$ carrier state elicited LTF response to $HB_sAg$. Majority of these subjects also exhibited LTF response to LSA. About 15% of $HB_sAg$ carriers who failed to manifest CMI to $HB_sAg$ showed significant LTF activity to LSA. Despite the differences in LTF responses to $HB_sAg$ and LSA in different population groups, the response to PHA was generally similar although it appeared to be somewhat depressed in subjects with acute hepatitis. These observations suggest that the induction of cellular responses against native liver specific proteins may be the underlying mechanism of liver damage during acute HBV infection. It is possible that the persistence of specifically sensitized clones of T-lymphocytes may lead to establishment of chronic liver disease.