924 UPTAKE OF RADIOACTIVE CALCIUM (^{45}Ca) BY NEUTROPHILS (PMN) FROM HUMAN NEONATES. Ronald G. Strauss and Esther L. Snyder. University of Iowa College of Medicine, Departments of Pathology and Pediatrics, Iowa City.

Abnormal chemotaxis, deformability, and lectin capping of neonatal PMN may be due to abnormal plasma membrane structure or function (eg, altered Ca uptake after perturbation). We studied ^{45}Ca uptake by blood PMN from 45 neonates. PMN and ^{45}Ca were incubated at 37°C . Basal ^{45}Ca uptake plateaued after 15 min and resting samples were taken. PMN were then stimulated; ^{45}Ca uptake increased for 15 min, and samples were taken. Samples were centrifuged through silicone oil with $> 99.9\%$ PMN and $<0.5\%$ free ^{45}Ca entering the pellet. Pellet radioactivity = PMN ^{45}Ca uptake. ^{45}Ca uptake (mean cpm) by resting PMN (N=45) was 1163 neonates and 1685 controls ($P < .0001$). ^{45}Ca uptake increased (2.2 - 145 fold) comparably ($P > .05$) in neonatal and control PMN after soluble stimuli (NaF and DMSO) or zymosan particles. Stimulated values (mean cpm) for neonates and controls were: NaF 113115 & 107421 (N=8); DMSO 164215 & 157944 (N=10); zymosan 2932 & 4380 (N=10). Neonatal PMN responded superiorly ($P < .02$) to FMLP; values were 4157 neonates vs 1297 controls. In conclusion, basal ^{45}Ca uptake by resting neonatal PMN was significantly decreased. The importance of this finding is undefined. Neonatal PMN increased ^{45}Ca uptake as well as or better than control PMN in response to several stimuli. Thus, abnormal membrane-dependent functions cannot be explained by failure of neonatal PMN to acquire extracellular calcium following stimulation.

925 PLATELET-VASCULAR ABNORMALITIES IN HEMOPHILIA A. M.J. Stuart, P.D. Sadowitz*, R.W. Walenga*, J.G. Kelton*. SUNY, Upstate Medical Center, Syracuse, N.Y. and McMaster University, Hamilton, Ontario.

A prolongation of the bleeding time (BT) occurs in hemophilia, although no cause has been elucidated. We evaluated B.Ts, plt. thromboxane B_2 (TXB_2), plasma $6\text{KPGF}_{1\alpha}$, serum immune complexes (I.Cs) and plt associated IgG (PAIgG) in 31 males with severe hemophilia A and in 17 controls. Compared to control values of $5.7 \pm 0.65'$ (mean $\pm$ 1SD) prolonged B.Ts were seen in 84% of patients (12.85 ± 4.74 ; $p < 0.001$). $\uparrow \text{TXB}_2$ with either thrombin (1.0 ± 0.67 pmol/ 10^5 plts) or arachidonic acid (AA) (4.16 ± 2.25) was observed in the hemophiliacs compared to controls (1.92 ± 0.81 and 6.23 ± 2.15 ; $p < 0.005$). All patients with $\uparrow \text{TXB}_2$ levels post thrombin had concomitant $\uparrow$ with AA, suggestive of a plt cyclooxygenase defect. $\uparrow \text{PAIgG}$ levels were seen in 72% of patients, and 70% were positive for I.Cs. No relationship between PAIgG, B.Ts and TXB_2 was noted. However 95% of hemophiliacs with B.Ts $> 10'$ were positive for I.Cs. Only 12% of patients negative for I.Cs. had B.Ts $> 10'$. A bimodal distribution of plasma $6\text{KPGF}_{1\alpha}$ levels was noted in the hemophiliacs when compared to controls. 8/9 patients with $\uparrow$ plasma $6\text{KPGF}_{1\alpha}$ levels were positive for I.Cs, while 4/5 patients with the highest titers of I.Cs had $6\text{KPGF}_{1\alpha}$ levels at or 2SD below controls suggestive of an "exhaustion" phenomenon. Initial stimulation of $6\text{KPGF}_{1\alpha}$ by I.Cs followed by vascular exhaustion would explain the bimodal distribution. Correlations between BT, TXB_2 and $6\text{KPGF}_{1\alpha}$ ($r = 0.74$) were noted. Our data suggests that the majority of hemophiliacs have prolonged B.Ts which correlate best with I.C. induced vascular injury.

926 TOWARD THE ELIMINATION OF VARICELLA IN CHILDREN WITH LEUKEMIA. Jean Taylor-Wiedeman, Philip A. Brunell, Clementina Geiser, and Ziad M. Shehab. U.T. Health Science Center, Department of Pediatrics, San Antonio, Texas.

Varicella may produce significant illness in children with leukemia. To reduce morbidity, Varicella Vaccine (VV) was given as soon as possible following diagnosis of leukemia. Twenty-eight of 29 given VV no earlier than 1 year post diagnosis seroconverted; 1 developed mild varicella at 1 year and another at 2 years post immunization. In addition, 5 of 28 lost antibody. Only 10 of 17 children immunized earlier than 1 year post diagnosis had satisfactory responses to VV. Passive immunization was started at the time of diagnosis of leukemia to provide protection prior to the time active immunization could be successfully accomplished. VZIG produced low levels of antibody that was detectable <10 -12 weeks. Patients who received transfusion of platelets, but not packed red blood cells, were found to have much higher antibody levels. Protection of children with leukemia can be achieved by giving VV one year post diagnosis. Prior to this time passive immunization, probably best accomplished with an intravenous product, could reduce morbidity from varicella in these high risk children.

927 EFFECTIVE TREATMENT OF FAMILIAL ERYTHROPHAGOCYTIC LYMPHOHISTIOCYTOSIS. Carol I. Teitlebaum, Lawrence D. Frenkel, David Senitzer (Spon. by Margaret G. Robinson). Department of Pediatrics, Medical College of Ohio, Medical College of Ohio Hospital, Toledo, Ohio.

Familial erythrophagocytic lymphohistiocytosis (FEL) is a rare condition with a life expectancy of 8 weeks from onset of illness. Successful treatment is unknown. A 4 week old male presenting with fever, jaundice, hepatosplenomegaly with liver failure, and pancytopenia was diagnosed as FEL on the basis of bone marrow, spleen, and liver biopsies. The tissues showed predominance of histiocytes and immunoblasts with hemophagocytosis. Viral etiology was excluded. Post mortem examination of the liver and spleen of a male sibling with a similar clinical course was identical with the findings in our patient. Supportive care with blood products, high dose decadron and splenectomy improved the clinical status minimally. Data suggested immunologic suppression with decreased T helper and increased T suppressor activity ($T_4=15.4$; $T_8=32.2$; $T_4/T_8=0.4$). Based on the premise that FEL represents dysfunction of T cell control of monocytes, the experimental drug Cyclosporin A (Sandoz) was used to control this apparent immunologic phenomenon. The patient is now surviving with normal hematologic and hepatic parameters at 10 months after diagnosis. There is no significant myelosuppression or renal toxicity using Cyclosporin A (10mg/kg/d) and prednisone (0.5mg/kg/d). Cerebral atrophy on CAT scan (demonstrated prior to experimental treatment) persists unchanged. Cyclosporin A appears to be an effective treatment of familial erythrophagocytic lymphohistiocytosis.

928 RED BLOOD CELL (RBC) GLYCOLYTIC ENZYMES AND GLUCOSE-6-P DEHYDROGENASE ACTIVITY IN PRE-TERM INFANTS ON THE FIRST DAY OF LIFE. Susan F. Travis, Savitri P. Kumar*, Linda M. Sacks*, and Maria Delivoria-Papadopoulos. Jefferson Med. Coll., Dept. of Pediat., Cardeza Fnd. for Hem. Res. and Univ. of Penn. Depts. of Pediat. & Physiol., Phila., PA.

Activities of RBC hexokinase, phosphofructokinase (PFK), aldolase, phosphoglycerate kinase (PGK), enolase (ENO), pyruvate kinase (PK) and glucose-6-P dehydrogenase (G-6-PD) were evaluated on the first day of life in 30 appropriate for gestational age preterm infants divided into 3 groups according to gestational age: 28-30 wks, 31-33 wks, and 34-36 wks and compared to prior results obtained in term infants. Mean activities of age-dependent RBC enzymes PK and G-6-PD were significantly higher than in term infants (PK: 512.1 ± 92.1 vs 385.6 ± 57.6 units/100ml RBC; G-6-P: 432.1 ± 42.2 vs 334.5 ± 38.7), suggestive of a younger RBC population in preterm infants. PGK activity was only slightly higher in preterm infants (454.3 ± 31.3 vs 441.3 ± 32.2 units), but mean ENO was significantly increased (560.2 ± 48.7 vs 346.7 ± 33.5 units/100ml RBC) when compared to term infants. RBC enzyme activity tended to increase as gestational ages of infants decreased but no statistical difference was noted in infants between 28-30 wks and 34-36 wks. PFK activity was higher (182.4 ± 21.5 vs 139.5 ± 13.9 units/100 ml RBC), not lower, in preterm than term infants and correlated significantly with PK activity ($p < 0.001$). Thus, young mean age of RBC population in preterm infants results in higher PFK activity than anticipated. PFK activity, however, was significantly decreased for the age of RBC population as documented by evaluation of a 24 wk old infant with severe hemolysis and very young red cell population. Except for ENO activity, it appears the activity of RBC enzymes in preterm infants does not reveal a more "fetal" pattern.

929 TRANSIENT NEUTROPENIA OF CHILDHOOD. R. Valiaveedan S. Rao, S. Miller, A. Brown. SUNY/Downstate Med. Ctr/Kings County Hosp. Brooklyn

While neutropenia ($\text{ANC} < 1500$) related to acute infections in children is well known, natural history and appropriate management are unclear. Clinical data on 22 children referred for neutropenia over a 13 mo. period were reviewed. Twenty patients were age 3 mos.-3 yrs. and 2 were age 11 yrs. $\text{ANC} < 500/\text{cmm}$ in 18 patients. All patients were febrile at some point in their illness; the majority (13) had respiratory tract infection. Infectious agents were isolated in 6 patients (S. pneumo. 2, H.flu. 1, E.coli 1, RSV 2). Neutropenia may have been drug-related in 5 patients (carbamazepine 1, chloramphenicol 2, penicillin 1, ampicillin 1). Bone marrow aspiration, performed in 14 patients, was normal in 7; seven patients demonstrated maturation arrest.

Recovery from neutropenia was documented in 20 patients; one child has remained neutropenic for 12 months and one was lost to follow-up. Ten recovered within 1 wk. of referral, whereas 10 remained neutropenic from 1 to 6 wks. Time to recovery could not be predicted based on any clinical or laboratory parameters available.

These cases of childhood neutropenia appear to have been infection-related and 20/21 patients recovered. Superimposed bacterial infection did not occur even with $\text{ANC} < 500/\text{cmm}$; empiric treatment with broad spectrum antibiotics was not instituted. Bone marrow evaluation, while useful in excluding infiltrative disorders and aplastic anemia, was not predictive of the rate of recovery.