884 GLYCOLYTIC ENZYMES IN PREMATURE INFANTS ON THE FIRST DAY OF LIFE. Susan F. Travis, Patricia L. O'Neal, Savitri P. Kumar, Maria Delivoria-Papadopoulos, Thos. Jeff. Univ., Dept. Ped. and Cardeza Fdn. and Univ. Penna. Med. School, Dept. Ped. and Physiol. Philadelphia, PA

Prior studies of RBC glycolytic enzymes in term infants on the first day of life have revealed elevated levels of phosphoglycerate kinase (PGK) and enolase (ENO) and decreased activity of phosphofructokinase (PFK) when compared to both RBC's from normal adults and subjects with a RBC population of a similar young mean cell age, as reflected in pyruvate kinase (PK) activity. In the present study, 25 premature infants were studied on the first day of life to determine whether this increase in RBC PGK and ENO and decrease in PFK activities are greater in the preterm infant and vary with gestational age. There were 3 groups: 9 infants 28-30 wks of age; 9 of 31-33 wks and 7 of 34-36 wks. The age-dependent enzymes PK, hexokinase and aldolase were higher in premature infants than term infants indicating a RBC population of a younger mean cell age. The mean activities of both PGK and ENO were also higher in the premature infants than in term infants but did not differ significantly between 28-30 wks to 34-36 wks. Mean PFK activity, however, was higher, not lower in premature infants than in term infants. In contrast to term infants, PFK activity correlated well with the age-dependent enzyme, PK ($r=0.73$; $p<0.001$). Thus, it appears that the young mean RBC population present in the premature infant on the first day of life significantly influences PFK activity, resulting in higher levels than those anticipated at such a young gestational age.

885 FREE ERYTHROCYTE PROTOPORPHYRIN IN SICKLE CELL ANEMIA. Elliott P. Vichinsky, Klara M. Kleman and Bertram H. Lubin*. Children's Hospital and Bruce Lyon Mem. Res. Lab., Oakland, CA.

We recently conducted a screening program to determine the incidence of iron deficiency in 66 patients with sickle cell disease. We found mean free erythrocyte protoporphyrin (FEP) values of 168 µg/dl RBC in 50 HbSS patients in contrast to 81 µg/dl RBC in 16 HbSC patients and 60 µg/dl RBC in 9 normals. Six of the pts with sickle cell disease had iron deficiency anemia documented by response to iron. None of the sickle cell patients had evidence for lead poisoning. FEP in iron deficient HbSS patients was 308 µg/dl RBC, and in iron sufficient HbSS patients was 153 µg/dl RBC. The correlation between FEP and retic count was excellent ($p<.001$). Reticulocyte rich fractions prepared by stractan density gradient separation from HbSS, hereditary spherocytosis, and pyruvate kinase deficient RBC samples had 40% higher FEP levels than reticulocyte poor fractions. Spectrofluorometric analysis of red cell protoporphyrin extracted in acetone-acetic acid revealed a four-fold increase in the ratio of protoporphyrin IX/zinc-protoporphyrin in reticulocyte rich fractions compared to normal or reticulocyte poor fractions. We found iron deficiency in 26% of non-transfused sickle cell patients. Our FEP results suggest that elevated reticulocyte counts contribute to high FEP values in HbSS patients and that the protoporphyrin distribution in reticulocytes differs from that in mature erythrocytes. We suggest that if FEP is used to screen for iron deficiency in HbSS patients that the FEP value be adjusted for the reticulocyte count. Supported by NHLB Grant No. HL 20985-2 from NIH.

886 PERINATAL LEUKAEMIA. Raj P. Warrier, Yaddanapudi Ravindranath, Susumu Inoue, Abbas Emami, Jeanne M. Lusher. Children's Hospital of Michigan, Detroit, Michigan, 48201.

Perinatal leukaemia is a rare disorder and extensive cytochemical, morphological and immunological studies have not been carried out in most of the earlier cases. We report four cases of perinatal leukaemia with clinical, morphological, cytochemical, immunological and cytogenetic correlations. Two had acute lymphoblastic leukaemia (ALL) and the other two had acute myeloblastic leukaemia (AML). The age at diagnosis was 5 hours, 9, 34 and 64 days. Initial hemoglobin values ranged from 5.5 gm/dl to 14 gm/dl. WBC counts varied from 60,000/mm^3 to 666,000/mm^3. In the two with ALL the lymphoblasts were B antigen positive null cell type (Blood 49:371, 1977). Three had normal karyotypes in the marrow cells before treatment. In the fourth a 46XX, 18 P- karyotype was noted with the father showing 46XY t (18p; 10p). Three patients died at 5, 47 and 223 days after diagnosis. One, who had AML, and who is the male member of a dizygotic pair of twins, is alive and disease free 14 years after diagnosis. Our observations indicate that the reported higher incidence of AML in neonates should be more carefully verified. Similar to ALL in somewhat older age groups of children, null cell B antigen +ve ALL appears to be the common phenotypic expression of ALL in neonates. Exchange transfusion was found to be useful in reducing the immediate risk of metabolic and leukostatic complications. In addition, the disease free survivor indicates that long term remission is possible in perinatal leukaemia.

887 INTRANASAL DDAVP IN PATIENTS WITH VON WILLEBRAND'S DISEASE AND HEMOPHILIA A. Indira A. Warrier, A. Samy Khalifa, and Jeanne M. Lusher, Children's Hospital of Michigan, Detroit, Michigan 48201.

The effect of a single intranasal dose (200 ugm) of 1-deamino-9-D-Arginine Vasopressin (DDAVP) was studied in 12 individuals with von Willebrand's disease (vWD) and 2 with moderate hemophilia A. Crossed immunoelectrophoresis of VIII:RAg demonstrated normal electrophoretic mobility in each of the vWD subjects. Components of the factor VIII system (VIII:C, VIII:RAg, VIII:R Cof) were assayed pre- and 90 and 180 minutes post-DDAVP. Each of 11 subjects with mild or moderate vWD had an increase in VIII:C activity (avg. 2X increase), 8 of 11 had an increase in VIII R:Cof, and 9 of 11 had an increase in VIII:RAg. The twelfth vWD subject, who had severe vWD, had no rise in any of these components. Of 4 vWD subjects who had pre- and post-DDAVP template bleeding times (BT) performed, the only one who had a prolonged baseline BT showed a normal BT 90 minutes post-DDAVP. One vWD subject who had demonstrated an increase in all F VIII components after DDAVP, later underwent dental extractions 90 minutes after DDAVP. No excessive bleeding was noted. Two individuals with moderate hemophilia A (baseline VIII:C values of 0.02 u/ml and 0.07 u/ml) were also studied. One had a rise in all components of the factor VIII system post-DDAVP while the other did not. We conclude that DDAVP results in transient improvement in selected individuals with vWD or moderate hemophilia A. This drug thus warrants further study as an alternative to blood components in the management of vWD and mild hemophilia.

888 ACUTE MYELOMONOCYTIC LEUKEMIA AS THE FIRST HEMATOLOGIC MANIFESTATION IN A PATIENT WITH FANCONI ANEMIA Michael Weiner, Kwame Yeboa, Dorothy Warburton, Arleen Auerbach (Spon. by James A. Wolff) Columbia Univ., College of Physicians & Surgeons, Dept. of Pediatrics and the Memorial Sloan-Kettering Cancer Center, Dept. of Cancer Genetics, New York, N.Y.

Chromosome instability and an increased sensitivity to DNA cross-linking alkylating agents appears to be present in patients with Fanconi anemia (FA). A 5½ year old white female with short stature and microcephaly developed acute myelomonocytic leukemia (AMML). Chromosome analysis of the bone marrow showed a karyotypically abnormal clone with rearrangements consistent with AMML, as well as an increased rate of spontaneous chromosome breakage. The diagnosis of FA was confirmed by finding 8.07 (normal 0-0.10) chromosome breaks per cell in lymphocytes cultured with the difunctional alkylating agent diepoxybutane. A complete bone marrow remission was induced with 1 course of cytosine arabinoside 100mg/M^2/day IV X 7 by continuous infusion and adriamycin 30mg/M^2/day IV X 3. Severe pancytopenia presumably secondary to heightened chromosomal instability of the normal regenerating bone marrow elements necessitated significant drug reduction of all subsequent anti-leukemia treatment. Despite minimal chemotherapy the patient remains in a complete remission. This patient's course illustrates three points: first, the clinical spectrum of FA is quite varied; second, AMML may be the first hematologic event in FA without prior therapy with either androgens or corticosteroids; third, treatment requires significant chemotherapy dosage modification.

889 ERYTHROCYTE PROTOPORPHYRIN (EP): THE RATIO OF FREE EP TO ZINC-COMPLEXED EP IN NORMAL CHILDREN. Ray Yip, Amos R. Deinard, Samuel Schwartz, Betty Stephenson. University of Minnesota, Dept. of Pediatrics and Medicine, Minneapolis.

Most of the EP in red cells is in a zinc-complexed form (ZEP) whereas the small remaining fraction is in the free state (FEP). Recently refined methods allow quantitative measurement of both fractions (FEP + ZEP = EP). The ratio of FEP/EP was determined for 853 children aged 6 mo to 12 yrs in whom FEP, ZEP, Hgb, and ferritin were determined (150 also had blood lead measured). The mean FEP/EP ratio for the entire group was 9.3% with a 95% range of 2.7 to 23.5%. The FEP/EP did not vary with age, Hgb or iron status (based on ferritin). The FEP/EP ratio varied only to a minor degree with elevation in total EP and with blood lead. Even though high FEP/EP has been observed in hemolytic conditions associated with reticulocytosis, in this study most children with high FEP/EP (> 25%) were apparently healthy and did not have evidence of increased red cell turnover or elevated total EP. These children with high FEP/EP ratio may represent a "normal" variant. Longitudinal follow up of some children showed the FEP/EP ratio to remain relatively constant. High FEP/EP was found to cluster in certain families, which suggests the possibility of a familial defect in conversion of FEP to ZEP.