834 HOW TO EXCHANGE TRANSFUSE PATIENTS WITH SICKLE CELL DISEASE K. Ackerman, C. Seaman, W. Shevchuk, S. Piomelli. Comp. Sickle Cell Ctr., Columbia U., NY.

Simple transfusion in sickle cell syndromes raises the hematocrit (Hct), while still large proportions of cells with sickling potential are in the circulation, with a dangerous increase in blood viscosity. In certain life-threatening situations, exchange transfusion is the only means to rapidly decrease the circulating sickle cells. Computations of the amount and type of blood to be used are often needed with great urgency. We have developed an algorithm for computer analysis of the various factors to be considered: patient's Hct, desired amount of remaining sickle cells, type of blood needed and rate of exchange. When the Hct is < 19%, exchange transfusion can be performed with packed RBCs; when the Hct is 19%-33%, an initial exchange with packed RBCs has to be followed by transfusion with packed cells diluted to a Hct of 40% with saline. In those rare patients with Hct > 33% it is also necessary to reduce the Hct by removing 8% of the blood volume. Using these measures, an exchange transfusion can be performed to reduce the concentration of sickle cells to any level, without raising the Hct above a set limit (usually 41%). We have also derived through differential analysis, simple formulas to determine the amount and type of blood to be used, with good approximation, using a hand-held calculator. The assumptions of these models were verified in 8 patients with sickle cell syndrome who were exchange-transfused in the last few months. This appears to be a needed and useful tool in the management of patients with sickle cell syndromes in life-threatening situations.

835 Physiologic Anemia of the Newborn: in vitro evaluation of inhibitors of erythropoiesis. P.A. de Alarcon, E.M. Mazur, L.A. Miceli, K. South. MIBH, Cooperstown, NY, University of Iowa, IA, Myriam Hospital, Providence, RI.

Decreased hemoglobin concentration occurs at six weeks of age. Its mechanism is unclear. We studied six infants at six weeks of age and their parents, with the in vitro plasma clot assay for erythroid progenitor cells CFU-E and BFU-E. Serum samples and peripheral blood (PB) mononuclear cells (MNC) were obtained by venipuncture. Bone marrow (BM) aspirate MNC were obtained from a normal adult volunteer. MNC were fractionated into T-cells and non-T-cells by sheep erythrocyte rosetting. Some PB-MNC from each baby were pre-incubated with: a) fresh plasma (FP), b) rabbit complement (RC), c) FP and baby's serum (BS) and d) RC and BS, for one hour at 37°C. All cultures contained 5 x 10^4 cells per micro-well. 1 x 10^4 T-cells were added to some experiments. The baby's PB-MNC produced a mean of 7±2 BFU-E colonies per clot, and 9.5±5 BFU-E when cultured in the presence of BS. Pre-incubation with FP, RC, FP+BS and RC+BS produced 5±1, 4±1, 5.5±2 and 4±2 BFU-E respectively. Adding baby's T-cells to his/her parents' PB-MNC increased the number of BFU-E from 11 to 19. BM-MNC produced a control of 250±10 CFU-E and 30±6 BFU-E per clot. In the presence of BS there were 219±30 CFU-E and 26±8 BFU-E. The addition of BS to cultures of baby's PB-MNC or the pre-incubation of these cells with BS and complement showed no significant inhibition of PB-BFU-E. The baby's T-cells increased the number of PB-BFU-E. Similarly, BM-CFU-E and BM-BFU-E were not inhibited by BS. T-cell suppression of erythropoiesis or serum inhibitors of erythropoiesis, either complement mediated or not, do not explain the physiologic anemia of the newborn.

836 Down's syndrome and increased mean corpuscular volume (MCV), mean platelet volume (MPV) and neutrophil alkaline phosphatase (NAP). P.A. de Alarcon, L.A. Miceli, E.M. Mazur and K. Smith. Pathfinder Village, Edmonston, NY, MIBH, Cooperstown, NY, University of Iowa, IA, and Myriam Hospital, Providence, R.I.

Down's syndrome is associated with multiple hematologic abnormalities. Newborns with Down's syndrome may develop a myelodysplastic syndrome akin to leukemia. The incidence of acute leukemia is increased in Down's syndrome. These serious hematologic complications of Down's syndrome may be the reflection of a steady state of compensated dysmyelopoiesis.

We studied 28 otherwise healthy individuals 10-25 years of age with Down's syndrome with three easily accessible hematologic parameters: MCV, MPV and NAP. The NAP was performed by the azo-dye method of Ackerman on peripheral blood smears and a numerical score given. MCV and MPV were measured by the Coulter counter S plus IV. The Down's syndrome population had a MCV of 94.1±3.9 fl ($\bar{x}$±SD) a MPV of 9.3±0.7 fl and an NAP score of 161±54. The normal controls for the Coulter counter model S plus IV in our labaratory are a MCV 87±7 and a MPV of 8.9±1.5. The NAP score in normals has a mean score of 100±50. Down's syndrome mean values for MCV, MPV and NAP differed from the mean of normal population in our laboratory ($p = <0.01$, <0.05 and <0.01 respectively). MCV, MPV and NAP are all increased in Down's syndrome.

Macrocytosis, both of platelets and red cells with disordered metabolism of white cells suggest a state of disordered hematopoiesis in individuals with Down's syndrome.

● **837** INFLUENCE OF Hb F AND α THALASSEMIA ON SEVERITY OF SICKLE CELL DISEASE IN SAUDI ARABIA. Baker Al-Awamy, Gulzar A. Niazi, Mohammad Al-Mouzan, Mahtab A. Naeem, Mohammad T. Altorki, King Faisal Univ., Eastern Province College of Medicine, Dammam, Saudi Arabia. (Spon. by Howard A. Pearson)

The Hb SS disease of Eastern Saudi Arabia has attracted international interest because of its comparatively "benign" course, usually attributed to high levels of Hb F found in these patients. α thalassemia, resulting in hypochromic microcytosis, is also common in this area. We determined the prevalence of Hb S and α thalassemia at birth by cord blood screening.

	AF	AF + Barts	ASF	ASF + Barts	FS	FS + Barts
Number	6076	535	507	113	60	9
%	83.1	7.3	6.9	1.5	0.8	0.1

The influence of Hb F and α thalassemia on clinical severity of Hb SS disease was assessed. 12 infants with Hb FS at birth were compared with 5 with Hb FS + Barts. At 6 and 12 months, levels of Hb F were identical in both groups. However, infants with Hb FS + Barts were microcytic and had less hepatosplenomegaly, infections and dactylitis. 8 Hb SS adults with microcytic RBC and normal Hb A_2 were compared to 8 with normocytic RBC. Although Hb F levels were similar ($\bar{x}$ 11.9% vs 12.8%), the microcytic group had few clinical symptoms and no hospitalizations. The normocytic patients had frequent painful crises and hospitalizations. We found no relation between levels of Hb F and clinical severity in Saudi infant and adult patients with Hb SS disease. Rather, hypochromic microcytosis appeared to be a more important predictor of less severe clinical disease.

838 FIVE YEARS' EXPERIENCE WITH CHRONIC TRANSFUSIONS IN SICKLE CELL ANEMIA (SSA) USING DONORS MATCHED FOR MINOR RBC ANTIGENS. D.R. Ambruso, J.H. Githens, L.J. Ruder, W.M. Vaughn, D.J. Dixon, R. Alcorn, T. Hays, Dept. of Ped., Univ. of Colo. School of Med., Colo. Sickle Cell Center, Bonfils Memorial Blood Center, Children's Hospital, Denver.

In 1978, a chronic transfusion program was initiated for selected SSA patients using donors closely matched for 17 RBC antigens (C,D,c,E,e,Kell,P,M,N,S,s,Fy^a,Fy^b,Jk^a,Jk^b,Le^a,Le^b) because isoimmunization is common in black patients receiving random donor transfusions (34% in our population). This is due to the high frequency of negativity for certain minor RBC antigens (e.g. C,E,Fy^a,Fy^b,Le^b) in blacks and the likelihood of positivity for these antigens in Caucasians. Twelve patients who had received multiple previous "unmatched" random donor transfusions were then given 1-3 units of "matched" RBCs every 3 weeks over a period of 7-70 mo. Each patient was matched as closely as possible with 8-30 donors selected from over 1000 genotyped blacks.

Using "Unmatched" Donors			Using "Matched" Donors		
Total Units	Pts. with Antibodies	Total # Antibodies	Total Units	Pts. with New Antibodies	Total # Antibodies
492	8/12	18	1347	3/12	4

With multiple "unmatched" donors, the risk of antibody development per transfusion was 3.7% in contrast to 0.3% with "matched" donors. Two of the 3 "matched" patients with new antibodies (C,Kell;C) had presented difficulties in the initial genotype. Thus, only 1 patient had 1 new antibody (K^A_p) when matched strictly by protocol. Our data shows that matching for certain minor RBC antigens reduces isoimmunization in SSA patients.

839 MEASUREMENT OF 2,3-DIPHOSPHOGLYCERATE (2,3-DPG) AND ADENOSINE TRIPHOSPHATE (ATP) IN STORED BLOOD BY ^{31}P NUCLEAR MAGNETIC RESONANCE (NMR) SPECTROSCOPY. D.R. Ambruso, B. Hawkins, D.L. Johnson, A.R. Fritzberg, and W.C. Klingensmith III, E.R.B. McCabe (Spon. by Wm. E. Hathaway). Univ. of Colo. Sch. of Med. and Bonfils Mem. Blood Ctr., Denver, CO, and the Regional NMR Ctr., Colo. State U., Ft. Collins, CO.

Adequate levels of ATP and 2,3-DPG in stored blood are important for in vivo survival and oxygen transport of red blood cells after transfusion. ^{31}P NMR spectroscopy allows noninvasive measurement of these compounds. In this study, biochemical assays, using coupled enzyme systems, and NMR spectroscopy performed in Nicolet 150 spectrometer operating 60.74 MH_z in the pulse-Fourier transform mode were used to quantitate levels of ATP and 2,3-DPG during a five-week storage period of both whole blood (WB) and packed red blood cells (RBCs). Blood was drawn into CPD-adenine and processed according to standard blood bank procedures. Results for WB are shown below expressed as μmol/gm Hgb (mean ± SEM).

Storage	ATP Levels		2,3 DPG Levels	
(Days)	NMR	Biochemical	NMR	Biochemical
1	4.10 ± 0.70	4.61 ± 1.38	12.65 ± 1.83	13.30 ± 0.97
22	4.40 ± 1.10	4.59 ± 0.50	1.36 ± 0.25	1.87 ± 0.84
36	3.95 ± 0.93	3.75 ± 0.69	0.82 ± 0.25	0.66 ± 0.18

Both techniques yielded similar results for ATP and 2,3-DPG with the expected decrease during storage. Results were similar for PRBC. NMR spectroscopy provides a noninvasive technique for quantitating ATP and 2,3-DPG which could be applied to evaluation of disease states and quality control of stored blood.