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MODIFICATION OF OXYGEN AFFINITY IN SICKLE CELL ANEMIA
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Polymer formation in sickle cells is dependent on the concentration of deoxygenated sickle hemoglobin. BW12C and BWA589C are aromatic benzaldehydes that cause a decrease in oxygen affinity through stabilization of the oxy-(R)-conformation. We studied the effect of these drugs using a gravity filtration technique that is sensitive to small amounts of intracellular polymerized hemoglobin. Pur. suspensions of Hb SS red cells were deoxygenated with nitrogen and filtered through polycarbonate membranes of 5 μm pore diameter. With progressive deoxygenation of the suspensions, an oxygen tension (saturation) is reached at which rapid loss of filterability occurs. This 'critical PO_2' correlates with the polymerization tendency of the sample.

Both agents reduced the critical PO_2 and modified the amount of Hb S in a dose dependent fashion. There was a significant correlation between the change in critical PO_2 and the amount of Hb S modified by both drugs. Neither agent had any effect on erythrocyte morphology or hemolysis. These agents have a potential therapeutic role in sickle cell disease.

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HEMORHEOLOGIC PARAMETERS IN CHILDREN WITH HOMOZYGOUS SICKLE CELL ANEMIA RECEIVING CHRONIC RED CELL TRANSFUSIONS FOR LARGE CEREBRAL VESSEL VASCULOPATHY (LCVV). H. Sabio, V. McKie, K.M. McKie, T.L. Jeraldo, R.A. Adams, and A.E. Felice. Medical College of Georgia, Augusta, Georgia.

Five children with homozygous sickle cell anemia (SCA) without associated alpha thalassemia receiving partial exchange transfusions designed to maintain the % Hb S between 30-35 for management of LCVV were studied. Whole blood viscosities were determined utilizing a mechanical impedance viscometer-Sonoclot R (90 sec^{-1}). Viscosity was determined in samples with hematocrits adjusted with autologous plasma to 40% (VwB_{40}). Erythrocyte centrifugal packing (ECP) and aggregation with polybrene were also determined. Four children receiving pre-operative partial exchange transfusions were also studied. In these patients, abnormal polybrene-induced erythrocyte aggregation and ECP were corrected with transfusions. In the chronically transfused erythrocyte aggregation and ECP remained abnormal. VW_{40} in the untransfused patients was significantly elevated. VW_{40} of patients who had received pre-operative transfusions or chronic transfusions was not different from normal. These findings suggest that a partially suppressive transfusion regime is clinically successful. Viscosity measurements appear useful in the monitoring of chronic transfusions in SCA.

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EM QUANTITATION OF HbS POLYMER IN SS RBCs AND RHEOLOGICAL CORRELATION by A. Anne Kaperonis, M.D., Robert G. King, Ph.D., Jeanne A. Smith, M.D. and Shu Chien, M.D., PH.D., Columbia Univ. Col. Physicians & Surgeons, New York, NY.

Flow abnormalities of blood in sickle cell (SS) anemia mainly result from polymerization of deoxygenated sickle hemoblogin (HbS) at low oxygen tension. In this investigation we attempted to quantitate the relationship between molecular changes of HbS with deoxygenation and the altered rheological behavior of the SS red cell (RBC) suspensions. Using a Weissenberg Rheogoniometer, the viscous (η') and elastic component (η'') of the complex modulus (η^*) were measured in oscillatory shear, and apparent viscosity in steady shear. Fixed SS RBCs were analyzed by quantitative stereology of electron micrographs and the degree of polymerization calculated from the volume fraction of the cells occupied by fiber (V_p). Increases in V_p resulted in attendant increases in both η' and η'' ($p < 0.001$). For a mean MCHC 35.7±1.6 of the 80% RBC suspensions examined, polymer was noted from an oxygen saturation of HbS below 85%. We conclude that the changes in the viscoelastic characteristics of the SS RBC suspensions are directly related to the extent of fiber formation, which in turn is related to the oxygen saturation of HbS.

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