●764 RECYCLING OF RETICULOENDOTHELIAL IRON IN VIVO IN THE ABSENCE OF APOTRANSFERRIN. Jamie E. Lamb*, Catherine M. Craven*, John H. Ward+, James P. Kushner+, Jerry Kaplan*. (Spon. by Harry Hill) University of Utah School of Medicine, University Hospital, Department of Pathology and Internal Medicine, Salt Lake City, UT.

Plasma iron is largely derived from macrophage recycling of iron from red blood cells (RBC). We hypothesize that iron-free transferrin (apoTf) is required for iron distribution to the erythron but is not obligatory for release of macrophage iron. We tested this hypothesis using a strain of mutant mice with atransferrinemia. These mice have serum Tf concentrations of 1% or less, suffer from iron-limited erythropoiesis but exhibit parenchymal organ iron-overload. We synthesized ^{59}Fe-methemalbumin, incorporated it in vitro into sheep RBC and coated the sheep RBC with IgG. Coated and uncoated sheep RBC were injected IV and IP respectively into both normal and atransferrinemic (Hpx) mice. At intervals the quantity of ^{59}Fe in the blood and selected tissues was determined. In normal mice the ^{59}Fe disappeared from blood with a t1/2 of 3.1 min. By 30 min. 20-30% of the injected counts were found in liver and spleen. By 72h, hepatic and splenic iron fell and 15% of the injected counts were detected in RBC. Hpx animals cleared injected sheep RBC and deposited ^{59}Fe in the liver and spleen in a similar way. By contrast, Hpx mice were unable to recycle iron to the erythron (<1%) but did recycle iron to other organs. By 1 wk, 6% of the injected counts were found in the acinar cells of the pancreas. These data indicate that apoTf is not required for recycling of RES iron to the parenchymal organs but is required for recycling to the erythron. Iron distribution in the Hpx mouse closely resembles that in children with congenital atransferrinemia.

†765 IDENTIFICATION OF A NEW STRUCTURAL VARIANT OF α SPECTRIN IN A DOUBLE HETEROZYGOUS FORM OF HEREDITARY PYROPOIKILOCYTOSIS (HPP). J Lawler, T Coetzer, VN Mankad, RB Moore and J Palek. Dept. of Pediatrics, University of South Alabama College of Medicine, Mobile, Al. and Dept. of Biomedical Research, Tufts University School of Medicine, Boston, Mass. (Spon. RC Boerth)

Studies of erythrocyte spectrin were performed in a patient with hemolytic anemia and her family to understand the biochemical and genetic basis of HPP. The proband had decreased stability of her red cells when exposed to 45°C, typical of HPP. Her cytoskeleton exhibited instability to shear stress. There was an increase in the amount of spectrin dimers in a crude 4°C spectrin extract (58% compared to control value of 6±4%) indicating a decrease in spectrin dimer-dimer association. A structural analysis of her spectrin α I domain by limited tryptic digestion revealed a decrease in the normal 80000 dalton fragment and an increase in fragments of 74000, 61000, 55000, 21000 and 16000 daltons. A 25% decrease, relative to control in the spectrin to band 3 ratio on SDS-PAGE demonstrated partial spectrin deficiency in the membrane. On a discontinuous Percoll-Renografin gradient, the patient exhibited the presence of cells at higher densities than that found with control cells. Both parents are asymptomatic carriers with increased amounts of spectrin dimers at levels that are intermediate between the control and the patient. The father's digest had increased amounts of the 74000 dalton peptide, while the mother's digest had increased amounts of the 61000, 55000, 21000 and 16000 dalton fragments. The biochemical and genetic basis of this severe HPP is probably a double heterozygous state, one of which is a previously undescribed molecular defect of spectrin.

766 THE DETECTION OF OCCULT HEMANGIOMAS PRODUCING THE KASABACH-MERRITT SYNDROME UTILIZING Tc-99m LABELED RED BLOOD CELLS. Wellington Loh, John H. Miller, Edward D. Gomperts. Sponsored by Robert L. Baehner. University of Southern California School of Medicine. Childrens Hospital of Los Angeles, Departments of Pediatrics and Radiology, Los Angeles, California.

The Kasabach-Merritt syndrome consists of a cavernous hemangioma with internal intravascular coagulation associated with a microangiopathic hemolytic anemia. In some patients, activation of clotting and fibrinolysis leads to thrombocytopenia and coagulopathy. Often these lesions are cutaneous and are readily detected by clinical evaluation. Rarely these lesions may be occult, involving the liver, spleen, or even a soft tissue mass simulating a malignant tumor. In those patients in whom the lesion is readily apparent, there is no need for a diagnostic procedure although angiography is occasionally utilized for confirmation. The use of Technetium (Tc)-99m labeled red blood cells (RBC) provides a total body survey in those individuals in whom an occult hemangioma is suspected and establishes the diagnosis in those patients in whom a cutaneous lesion is present. The patient's own red cells are labeled in vitro and reinjected allowing excellent imaging. This procedure is relatively noninvasive and has a low radiation burden. We report the use of this technique in five patients presenting with Kasabach-Merritt syndrome. Three of these patients had occult or unsuspected lesions. No other imaging modality provides this graphic demonstration of these lesions. The technique of this procedure and illustrative cases will be presented.

●767 EVALUATION OF THE NEED TO IMMUNIZE CHILDREN WITH SICKLE CELL DISEASE WITH HEMOPHILUS INFLUENZAE TYPE B (HIB) VACCINE. Genevieve A. Losonsky, Ruth E. Luddy, Allen D. Schwarz, and John B. Robbins. University of Maryland, Department of Pediatrics, Baltimore, and The National Institutes of Health, Bethesda.

Insufficient data are available in which to base recommendations for the use of HIB polysaccharide vaccine in children older than 18-24 months who have functional asplenia who may be at increased risk for HIB systemic disease. Therefore, using a radioimmunoassay for detection of antibodies to the capsular polysaccharide of HIB, polyribosylribitolphosphate (PRP), we determined the PRP specific antibody levels in 36 children, age 2-20 years (mean 8.9 years) with SS or SC disease cared for in our hematology clinic to assess their potential risk for acquiring HIB disease. Although a precise protective level of PRP antibody has not been established with certainty, we used a level of 1 ug/ml to determine risk based on results generated in the Finnish field trial correlating this level with short term clinical protection.

20/36 (56%) of the children evaluated had PRP antibody levels less than 1 ug/ml. 15 of these "at risk" children (75%) were ≥ 5 years of age, with 8 children being 10 years of age or older. In fact, 10/20 (50%) "at risk" children had PRP antibody levels < 0.25 ug/ml with 50% of these being older than 5 years of age. Based on these data showing low levels of PRP antibody in older children with hemoglobinopathies, recommendations for administration of HIB vaccine should be extended to all children with such chronic conditions.

768 COULTER S+IV THREE PART DIFFERENTIAL SCREEN IN SELECTED PEDIATRIC PATIENTS. Naomi L.C. Luban, Ghiselaine Gautier, Lisa A. Kammerman (Spon. by G. Rosenquist). George Washington University School of Medicine, Children's Hospital National Medical Center, Department of Laboratory Medicine, Wash. D.C.

Using descrimination of WBC volumes, the Coulter S+IV provides an automated three part differential (3PD). The validity of the 3PD is checked by computer via a system of algorithms that confirm the shape of the WBC volume histograms and flag the abnormalities into R-flag, (abnormal WBC volume distribution) and Backlight flag, (high WBC count, abnormal size distribution or mononuclear count > 1.5x10^3/ul). We compared the 3PD in 525 samples from children age 1 mo. to 24 yrs. to those obtained by manual counting of 100 WBC cells. Samples were collected by fingerstick (n=109) or by vein (n=402) from a)same day surgery (n=162); b)general pediatric medical clinic (GPMC) (n=156); c)cardiology clinic (n=43); and d)hematology-oncology clinic (n=164). Specifically excluded were specimens from neonates and in-patients. The relationships between the manual and automated 3PD for S+IV using simple linear regression for venous samples was lymph r=.922, gran r=.919, mono r=.197, and for capillary samples was lymph r=.936, gran r=.914, mono r=.315. 49.9% of all specimens were flagged: 42.8% R flag + 7.1% backlight flag. The most frequent % flagging occured in the capillary specimens (63%) and in patients from hem/onc (55.8%). The least amount of flagging was in cardiology (29.5%) and GPMC (30.3%) regardless of collection method. Current Coulter methodology makes the 3PD useful for only a small subsection of routine outpatient differentials.

769 CHOLECYSTECTOMY IS SAFE IN SICKLE CELL ANEMIA. Betty Malone, Steven Werlin, Department of Ped., Medical College of Wisconsin, Milwaukee, Wisconsin.

Although recurrent abdominal pain due to gallstones is common in children with sickle cell anemia, cholecystectomy is recommended reluctantly because complications may be as high as 37%. We performed elective cholecystectomy in 12 children (8 M, 4 F; ages 4-19 yrs, mean 11.2 yrs) with abdominal pain related to gallstones in 10 and acute cholecystitis in two. Seven patients had jaundice, 6 nausea, 5 fat intolerance, and 3 biliary colic. Seven children were prospectively studied. Two simple transfusions (10 ml/kg PRBC), designed to decrease HgbS to 30% and to increase total Hgb to 10 gms%, were given preoperatively 2-3 weeks apart. Hgb electrophoresis was rechecked after the second transfusion. A third transfusion was given the day before surgery. The charts of 5 additional patients who received less intense preoperative transfusions were also reviewed. Preoperatively mean HgbS was decreased from 88% to 31% (range 7-38%) and mean Hgb was raised to 12.2 gms% (range 9.5-14.5 gms%). There were no intraoperative complications. Four patients had elective appendectomy and 2 umbilical hernia repair. Although no patients developed complications related to sickle cell anemia, one had ileus for 3 days and one had bigeminy requiring lidocaine. Hospitalization was brief (average 6.3 days, range 4-8 days). At 6 month followup, abdominal pain had resolved in all patients except one who developed pancreatitis. CONCLUSIONS: With proper preoperative transfusions, elective cholecystectomy is safe in children with sickle cell anemia.